Motor Assessments in Cerebral Palsy:

A study of the mechanisms of equinus, the functional neuromuscular angle, clonus, alternating movements and posture.

by

Jean-Pierre Lin

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The epidemiology of cerebral palsy (CP) is reviewed followed by an outline of the mechanisms of hypertonus. Mechanisms and clinical correlates of dynamic and passive equinus were studied in hemiplegic cerebral palsy which could not be explained solely in terms of muscle weakness or imbalance of power at the ankle or spasticity.Peripheral muscular transformation and increasing intrinsic stiffness with abnormal postures and muscle activation patterns are more likely explanations: a developmental model of equinus is advanced.

Electromyographically quiescent muscular creep in hemiplegia is demonstrated in muscles that might have been classed as "tonically spastic', along with muscle moulding and stress-relaxation, confirming a peripheral transformation of the plastic or time-dependent properties of muscle following long-standing cerebral injury. These mechanisms are independent of reflex or supraspinal electrical 'tonus': the success of some physical therapies (stretches, orthotics and serial plastering) may depend in part on the properties of muscular creep and stress-relaxation.

Reflex excitability of muscle acting at the knee and ankle was studied using manual discontinuous ramp or rhythmic sinusoidal stretches. Excitability of hamstrings and quadriceps muscles was greatest close to maximum knee extension, but grouped hemiparetic and nonparetic muscles behaved similarly. At the ankle, sinusoidal stretches allowed more graded reflex quantitation than ramp stretches. No muscles were excited at walking speeds of sinusoidal stretch. For more than half the cases, excitability was similar in hemiparetic and nonparetic limbs: in the remainder, the nonparetic reflex threshold was not reached. The surface EMG relation to standing, isometric and isotonic tasks is explored with respect to fine motor dexterity at the ankles and toes.

The influence of the joint angle on stretch reflex excitability of the soleus muscle at the ankle has been studied in children and adults. For all subjects, reflex EMG and mechanical twitch torque gain were trivial at *resting plantarflexion*. The *reflex EMG gain* reached a maximum between -15° and -10° of plantarflexion beyond the neutral angle, 0°, defined as the foot at right angle to the tibia, diminishing steeply with further dorsiflexion. The *reflex mechanical gain* rose to a peak between 0° and +10° of dorsiflexion beyond neutral, declining steeply thereafter. By contrast, axonally stimulated muscle twitch torque increased serially up to +30° dorsiflexion beyond neutral. The reflex twitch time increases with

dorsiflexion, rising from a mean of 277ms at -25° and 285ms at neutral to 370ms at +35°: a 33.5% increase. The in *vivo* excitability of the spinal alpha-motorneurone pool is strongly influenced by muscle length, giving an optimal neuro-mechanical angle +/-10° about neutral,which is a source of inter- and intra-subject variability if the joint angle is not controlled. Soleus muscle twitch characteristics showed a five- to eight-fold increase in peak force associated with a ten-fold reduction in compliance in the first two decades: apparently speeding up in the first decade. The slowest twitches occurred in the youngest children. Similar results were obtained in congenital hemiparetic CP. Heterogeneity of the hemiparetic data is evident when compared to nonparetic and control data with clear differences in the clonic (fast twitch) as opposed to nonclonic (slow twitch) muscles. In four cases with clinical clonus, the clonus frequency was reduced by passive dorsiflexion. Plaster immobilisation for one month brought out clonus which was previously absent in a further case, causing a fast-twitch soleus phenotype to emerge in this and another case immobilised for six weeks. Follow-up after heelcord lengthening in one case showed that clonus frequency decreased as the muscle twitch strengthened and slowed. Short and long-term peripheral therapies appear to regulate neuromuscular excitability according to whether muscles are loaded or unloaded.

The central and peripheral factors regulating the physiology of alternating movements (dexterity) are examined in the light of the foregoing: muscle maturation and use of the optimal neuromuscular joint angle are postulated as factors contributing to a doubling in speed in the first decade and a sub-optimal joint angle is demonstrated in 'paretic' states.

The influence of tonic labyrinthine reflexes on posture in diplegia of prematurity and the phenomenon of spontaneous extensor toes has been demonstrated together with its abolition by sleep. Implications for CP classification and selection for treatment (s), with particular emphasis on the inappropriate selection of 'antispastic' treatments, is discussed. A clinical distinction between the phenomena of abnormal postures and spasticity is advanced.

Summary. Peripheral constraints on the motor manifestations of CP have been demonstrated along with short and long-term adaptations to physical interventions leading to a thesis of optimal functional muscle lengths and postures.

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1. Cerebral Palsy

This section attempts to clarify the clinical phenomenology that are the cerebral palsies and to understand the current state of knowledge regarding natural history, the indications and likely benefits and complications of treatment.

1.1 Definition of Cerebral Palsy

Cerebral palsy (CP) is neither an illness nor a disease but is a medical syndrome or collection of symptoms and signs more or less arbitrarily grouped into distinct entities. The term "cerebral palsy" has been defined as:

> "..a non-progressive disorder of movement and posture due to a defect or lesion of the immature brain.." (Bax 1964)

The term cerebral palsy thus refers to the motor manifestations of damage or anomalies of the developing brain. Damage may arise by any means (eg genetic defects, migrational defects, cerebral malformation, hypoxic-ischaemic encephalopathy (HIE), trauma, infection, inflammation, haemorrhage, infarction, non-infarctive white matter disease) but the damaging process is thought to be static in contrast to the progressive neurological disorders which continue to damage the brain. In the absence of extremely severe initial damage, the brain continues to grow and develop, often at a slower rate than normal.This continued growth and development, coupled with physical growth, produces a changing clinical picture In addition to excluding conditions of progressive neurological damage, the term "cerebral palsy" usually excludes disorders of the spinal cord, peripheral nerves, neuromuscular junction and muscles (see below for a further discussion). The motor manifestations include both delayed development and abnormal function of the voluntary motor system including gross motor and fine motor function, feeding, speech and eye movements. Epilepsy, disorders of behaviour, learning and cognition, including disorders of vision and hearing are complications of the same injuring process which produced the original motor disorder of cerebral palsy. Typically, this early brain damage produces abnormalities of muscle tone, reflexes, posture and movement control. Only these motor manifestations will be dealt with in the following sections.

The last decade has witnessed a growing interest in the mechanisms of cerebral palsy and there has been a drive towards adopting a consensual approach for case definition

with a view to making valid comparisons of the prevalence rates within and between populations. Despite this, Mutch and colleagues (1992) in a review of the subject given the title "Cerebral palsy epidemiology: where are we now and where are we going?" raise some important basic questions. Principal among these are:

> "1. How do you define a case? 2. How severe does it have to be to be counted. 3. How do you describe the condition so that everyone knows what you mean?"
>
> *Mutch and colleagues (1992)*

Mutch et al (1992) go on to define cerebral palsy as :

> "an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development."

A further inherent difficulty is acknowledged by the group:

> "There is currently neither an objective test that can be used to 'screen' for the cerebral palsies, nor a diagnostic test that can be used as a 'gold standard'. Reported prevalence depends on the use of an arbitrary level of functional disability, and it has been shown that there is substantial between- and within-observer diagnostic variability, both with regard to what is a case and to what type of case it is. In order to establish any validity in comparative studies we must develop a standard descriptive system, avoiding the use of ambiguous technical terms. The need for standard definitions has been made the more urgent, since the neurological signs, functional impairments and disabilities seen increasingly in the extremely preterm infant do not conform readily to patterns seen previously."
>
> *Mutch and colleagues (1992)*

The purpose of the rest of this section on cerebral palsy is to indicate that the "conditions", albeit changing, are likely to present a medical challenge for the foreseeable future; that they differ to a very large extent from the patterns of neurological impairments encountered in adult practice; that the case definitions previously and currently in use may, through lack of definition lead to misunderstandings in the likely mode of action of established and newer treatments; finally, that only an understanding of the pathophysiology of the motor disorders is likely to result in appropriate treatments.

1.2 Incidence and aetiopathogenesis

Determinations of the incidence of cerebral palsy are complicated by the fact that there is a continuously changing relationship between the brain lesion (s) and the functional impairment. The diagnosis may not be clear until the end of the first or second year. In the Collaborative Perinatal Project, two thirds of the children with "spastic diplegia" and half of all children with signs of "cerebral palsy" at their first birthday outgrew their symptoms by the age

of seven years (Nelson and Ellenberg, 1982). Accordingly, it is often several years before data on the incidence of CP in a given birth cohort can be ascertained.

The changing motor patterns and signs within the same child over time reflect underlying neuronal maturation, synaptic connectivity, axonal myelination and somatic growth:these stages of brain development and maturation in relation to the ontogeny of postures in the foetus and infant has recently been extensively reviewed (see Brown *et al*, 1997).

A further complication is that some very slowly progressive disorders may mistakenly be taken for CP so that unexpected deteriorations in a child's neurological condition should always lead to a questioning of the original diagnosis.

The overall incidence data varies from 2-2.5/1000 live births (Stanley and Blair, 1991, Mutch *et al*, 1992). Prospective studies , which include mild CP cases, have a higher estimate of prevalence than service registers, which only tend to see severely affected children. Despite changes in obstetric care practices including the identification of at-risk mothers, and the establishment of neonatal units for the care of the newborn, the incidence of CP has changed little over the past 30 years, pointing to non-obstetrical causes for CP.

Although the premature infant is 50-100 times more at risk of developing CP than his term counterpart, and there is little doubt that the absolute incidence of CP in the lowest birth-weight groups has increased in parallel with the increased survival of such infants (Stanley and Blair, 1991, Mutch et al 1992, Scottish Low Birthweight Study Group 1992), the relative incidence of cerebral palsy remains unchanged, since it is also the case that many more low birth-weight infants survive without motor disability (Stanley and Blair, 1991; Forfar *et al*, 1994). Stanley and Blair (1991) have estimated that for every preterm graduate of the neonatal unit who survives with disability, twelve appear to survive intact (table 1.2.1). This also suggests that neonatal intensive care (ITU) does not "produce" brain damaged infants, although the ratio of intact survivors to graduates with cerebral palsy is narrowing.

Modern accounts of cerebral palsy(CP) begin with the attribution of CP to difficult deliveries by William Little (1843) ie to perinatal causes. Fifty years later, Freud (1897) speculated that CP could represent the effects of "deeper-lying influences" on the development of the foetus ie antenatal causes. William Osler (1889) reviewed the nineteenth century literature on the subject and compared this with the case histories of 161 cases in

Table 1.2.1 Hypothetical numbers of survivors with and without cerebral palsy before and after neonatal intensive care for live born infants with a birthweight of less than 1500g, based on Western Australian data

	Before Neonatal ITU 1968-1971 rate per 100	After Neonatal ITU 1982-1985 rate per 1000	Difference
N° who survived 28 days	325	725	+400
N° with CP at 5 years of age	14	55	+41
N° of survivors without CP	311	670	+359
Ratio of survivors without CP:with CP	22.2	12.1	

after Stanley and Blair 1991

Philadelphia. Of these, 74.5% were hemiplegic, just over 12.4% had a double hemiplegia and only 6.8% had a paraplegia or diplegia. This contrasts with 44-74% with diplegia (depending on birthweight) in current-day figures (Hagberg *et al*, 1993). In Osler's day, acquired major childhood infections such as scarlet fever, whooping cough and other acquired fevers resulting in childhood encephalopathies (with fits and coma) in the first three years of life played an important role in the aetiology of CP. For an up to date review of pathogenetic mechanisms see Kuban and Leviton (1994). The pathophysiology of brain damage in CP remains poorly understood, but for the majority of term infants (born after 37 weeks gestation) who later develop CP, there is no identifiable birth injury or evidence of hypoxic-ischaemic insult (Blair and Stanley, 1988, Stanley and Blair, 1991).

In considering the pathogenesis of cerebral palsy it is important to consider the risk factors for a child being born with brain damage. It is usual to divide the risk factors into those likely to affect **a.** term, **b.** preterm and low birthweight infants, as well as to consider whether the damage affects the cortex, the white matter, the basal ganglia the cerebellum or a combination of some or all of these structures. It is possible, with neuroimaging, to characterise the lesions according to the putative mode of injury, principally destructive / necrotic (as shown in figures 1.3.1, 1.3.2 and 1.3.3) or non-infarctive in which the principal observable change is atrophy of the white matter.

a. Term Infants. It has been customary to look for risk factors for "birth asphyxia" in children with CP (see table 1.2.2). However it is now increasingly recognised that "birth asphyxia" as

defined by a conspicuous series of clinical events relating to an obstetric complication with established foetal distress, clinical evidence of asphyxia requiring resuscitation at birth and a neonatal syndrome of hypoxic-ischaemic encephalopathy and multi-organ failure (ie renal, liver and pulmonary disease) occurs in only a minority, 8-25%, of cases of children subsequently shown to have cerebral palsy (Blair, 1988; Torfs *et al*, 1990; see also Bax and Nelson, 1993 for a research definition of birth asphyxia). A study of 19,044 children born to mothers with monitored pregnancies and followed medically for 5 years demonstrated that 78% (31/42) of the children who developed CP did not experience birth asphyxia (Torfs *et al*, 1990).

Table 1.2.2. Risk factors for birth asphyxia.

1. Short or long birth spacing
2. History of spontaneous abortions / stillbirth
3. Thyroid hormones or oestrogen replacement in pregnancy
4. Twin pregnancy
5. Malpresentation
6. Postmaturity: ie >42 weeks gestation, "senile placenta"
7. Low Apgar scores
8. Abnormal foetal heart rate during labour
9. Congenital anomalies

after Cohen and Duffner 1981

(Though 95% of infants with these risk factors do not have cerebral palsy)

Paradoxically, the benefit of electronic foetal monitoring during labour has been called into question when it was shown to confer no benefit over intermittent foetal auscultation in terms of the prevention of birth asphyxia or the subsequent incidence of cerebral palsy in a now famous Dublin study (Grant *et al*, 1989). In an earlier revealing study, blinded assessors reviewing antenatal records found that the mothers of normal children had experienced worse intrapartum care than those whose children were known to have cerebral palsy.

b. Prematurity and low birthweight.

The preterm infant seems particularly at risk of cerebral damage, and the following can only briefly review some of the most important mechanisms, chief amongst which are intraventricular haemorrhage (IVH) and more importantly, periventricular leucomalacia (PVL).

The Scottish Low Birthweight Study Group (1992) identified 896 children born with birthweights less than 1750g, of which 217 died in the neonatal period, 36 died in the first year, 6 died between 1-2years and one died between 2-4years of life. Of the 636 survivors, 25 could not be assessed and the group were able to establish a neuromotor impairment in

82/611 (13.4%) survivors available for assessment. The lower the birth weight, the higher risk of cerebral palsy, the incidence being 216/1000 (BWt<1000g), 141/1000 (BWt 1000-1499g) and 107/1000 (BWt 1500-1749g). A comparison with other regions or countries show similar results (Table 1.2.3):

Table 1.2.3.

International comparison of cerebral palsy rates per 1000 neonatal survivors; perinatal mortality rates (PNM) and birthweight distribution.

	1975-1978			1979-1982			1983-1984		
Birthweight (g)	PNM	%LB	CP/1000	PNM	%LB	CP/1000	PNM	%LB	CP/1000
Sweden									
<1500	511.5	0.5	58.3	371.0	0.6	93.3			
1500-2499	69.9	3.6	17.2	47.6	3.5	12.6	----------		
>2500	3.4	95.9	1.2	2.7	96.0	1.3			
W. Australia									
<1500	657.3	0.8	28.4	534.6	0.8	46.1	440.7	0.9	76.4
1500-2499	69.7	4.6	12.6	55.4	4.7	9.2	46.9	4.9	7.3
>2500	3.4	95.9	1.2	2.7	96.0	1.3	3.9	94.1	1.4
Mersey (UK)									
<1500	608.6	0.6	19.0	479.7	0.8	90.8	377.7	0.5	81.9
1500-2499	83.6	4.8	10.4	53.3	5.8	9.4	41.8	5.8	9.2
>2500	9.5	94.5	1.0	6.8	93.5	1.1	3.4	93.8	1.0

%LB= Birthweight distribution percentage for livebirths of known weight.
CP/1000= cerebral palsy cases per 1000 neonatal survivors.
Sources: Sweden G Hagberg; W. Australia L Watson and F Stanley; Mersey (UK) P Pharoah.
after Mutch et al 1992

Because of the fragility of the capillary bed in the periventricular germinal matrix of the preterm infant, intraventricular haemorrhage and post haemorrhagic hydrocephalus is more common in the preterm newborn (see Papille 1980, Volpe 1987, and Lin *et al*, 1992,1993, Vohr 1992). The neurological sequelae of severe (grades III and IV) IVH have been studied in relation to maximum intracranial pressure at the time of ventriculo-peritoneal shunting (Lin *et al*, 1992) and the clinical phenotypes reviewed (Lin *et al*, 1993).

Changes in the white matter of the brain such as periventricular leucomalacia (PVL) correlate better with the risk of CP than do intraventricular haemorrhages (IVH), see Levene, 1990. Up to 22-100% of preterm infants with PVL grow up to have cerebral palsy, and this rate increases if there are cysts in the white matter and especially if these are large, posterior and bilateral (see Kuban and Leviton, 1994). Table 1.2.4. shows the risk factors for PVL:

Table 1.2.4. Risk factors for PVL.

1. Placental vascular anastomoses
2. Twin gestation
3. Antepartum Haemorrhage and abruption
4. Inflammation of the umbilical cord or membranes: ie amnionitis
5. Low gestational age
6. Acidosis, low Apgars or asphyxia
7. Intracranial haemorrhage
8. Hypotension
9. Patent ductus arteriosus (PDA)
10. Sepsis
11. Necrotizing enterocolitis or surgery

after Kuban and Leviton ,1994.

A relatively new hypothesis is that in-utero infections such as chorioamnionitis trigger the onset of premature labour and cause cerebral damage with the release of destructive cytokines, eg tumour necrosis factor alpha (Adinolfi 1993; Leviton 1993). However, Christensen and Melchior (1967) in their book: "*Cerebral palsy: a clinical and neuropathological study* "quote the findings by Globus (1921) and Eisenstein and Taylor (1941) pointing to intrauterine inflammation with subsequent injury to the circulation as a cause for porencephaly.

The role of maternal infection is supported by a case control study from the Oxford Perinatal Epidemiology Unit involving 59 babies from a population-based register who developed cerebral palsy and 234 randomly selected controls. Cerebral palsy decreased with increasing gestational age and birthweight but increased with evidence of chorioamnionitis, prolonged rupture of membranes, and maternal infection (Murphy *et al*, 1995). The apparent relationship between maternal infection and subsequent risk of having a child with CP is shown in Table 1.2.5.

These recent studies indicate that maternal infection and chorioamnionitis may be two to four and half times as common in the mothers of children who subsequently present with cerebral palsy. Goldenberg and Andrews (1996) review the evidence for asymptomatic bacterial vaginosis and chronic endometrial infection prior to fertilisation as a cause for repeated preterm labour brought about by the endometrial decidua producing inflammatory cytokines which precipitate premature delivery.

More recently, Zupan *et al* (1996) have looked at the incidence of periventricular

leucomalacia (PVL) in 753 preterm infants born between 24 and 32 weeks gestation, admitted to intensive care and surviving at least 7 days.

Table 1.2.5. Maternal infection and risk of cerebral palsy

Author	Date	Variable	CP	(%)	Controls	(%)
Murphy *et al*	1995	Prolonged Rupture of Membrane	25/59	(42)	64/234	(27)
		Maternal Infection	22/59	(33)	40/234	(17)
		Chorioam-nionitis	10/59	(10)	8/234	(3)
		All causes	32/59	(43)	48/234	(20)
Grether *et al*	1996	Neonatal Seizures + chorioam-nionitis	6/42	(14)	0/72	(---)
Nelson *et al*	1996	Maternal Infection	14/78	(18)	13/300	(4.3)

They found that 69/753 (9.2%) of their preterm cohort had cystic PVL, being highest in babies born at 28 weeks (14%), the distribution according to gestational age is shown in table 1.2.6.

Table 1.2.6. Incidence of cystic PVL according to gestational age (GA)

GA (wks)	N	Early Death*	Survivors	PVL	Incidence of PVL**
<27	89	20	69	5	7.2
27	75	5	70	9	12.9
28	99	10	89	14	15.7
29	110	5	105	11	10.5
30	128	7	121	15	12.4
31	115	0	115	7	6.5
32	186	2	184	8	4.3
Total	802	49	753	69	9.2

*within the first seven postnatal days . **Calculated for infants surviving at least 7 days.

after Zupan 1996

Inflammatory prenatal events during the last days or weeks before the delivery were strongly correlated with the occurrence of PVL. Out of 144 cases of premature rupture of membranes (PROM), 25 cases (17.3%) developed PVL which compares with 38/202 cases (18%) of PVL where intrauterine infection was positively identified, but the percentage of cases with PVL rose to over 22% (22/98 cases) when PROM and intrauterine infection occurred together.The collective evidence points to the genuine role which maternal infections play in the phenomenon of preterm birth, PVL and cerebral palsy.

Murphy and colleagues (1995) were able to demonstrate that children born to pre-eclamptic mothers have a lower rate of CP (6/59:10%) than the control cases (56/234: 24%,). This was confirmed by Zupan *et al* (1996) who found that infants who had experienced chronic foetal stress with severe intrauterine growth retardation and those whose mothers had toxaemia of pregnancy (excluding eclampsia and placental abruption) seldom developed cystic PVL., the relative risk being less than 2% as compared to 22% following chorioamnionitis.

The issue of the "favourable v hostile intrauterine environment" is made more intriguing by the findings of Nelson and Grether (1995), who, in a recent case control study, demonstrated the apparent protective effect of magnesium sulphate (MGSO4) administered as an antihypertensive agent in pregnancy induced hypertension. They identified very low birth weight (VLBW) singleton infants (<1500g) surviving to three years with moderate or severe congenital CP from a cohort of 155,636 children born between 1983 and 1985 in four California counties and compared these with randomly selected VLBW control survivors with respect to whether or not the mothers had received MgSO4 to prevent convulsions in pre-eclampsia or as a tocolytic agent. This study showed that only 7.1% of the 42 VLBW infants with CP compared to 36% of the 75 VLBW controls were exposed to MgSO4, and this result was highly significant (Odds ratio 0.14, 95% confidence intervals 0.05-0.51).

This data coincides with the recently published data from the multicentre Eclampsia Trial Collaborative Group (1995) which showed that out of 1687 women recruited with eclampsia, data was available in 1680 cases of whom 453 were allocated MgSO4 versus 452 given diazepam and 387 allocated to treatment with phenytoin. The women given MgSO4 had a 52% lower risk of recurrent convulsions (95% confidence intervals 64% to 37% reduction in fits) compared with those given diazepam to control their fits and the MgSO4

group also had a 67% reduction in fits compared with the phenytoin treated group of eclamptic women.

The study by Nelson and Grether (1995) indicates that therapeutic and prophylactic MgSO4 trials may reduce the rates of cerebral palsy, but this has to be tempered by the confounding protection which pregnancy induced hypertension confers on the foetus (Murphy et al 1995, Zupan et al 1996).

1.3 Classification of Cerebral Palsy

Table 1.3.1 indicates a variety of ways of classifying the cerebral palsies.

Table 1.3.1 Schemes for Classifying the Cerebral Palsies.

i) Type of brain injury:	genetic, malformation, deformation, destruction (trauma/compression), infective, inflammatory, infarctive, haemorrhagic, non-infarctive periventricular leucomalacia (PVL)
ii) Timing of brain injury:	1st, 2nd, 3rd trimester, perinatal, postnatal.
iii) Site of brain injury:	cortical, cortical-subcortical, white matter , basal ganglia, brainstem, cerebellar or global
iv) Topography of signs:	monoplegia, diplegia, triplegia, quadriplegia, double hemiplegia
v) Motor manifestations:	hypotonic, ataxic, spastic, dystonic, dyskinetic
vi) Functional impact:	none, mild, moderate, severe
	impairment: disturbance at organ level
	disability: the consequence of impairment for function and activity
	handicap: the disadvantage to the individual arising from impairment and disability
	(*WHO International classification of impairments, disabilities and handicaps. Geneva , WHO 1980*)

But as indicated in the discussion of the definition of CP, Mutch and colleagues (1992) have suggested the adoption of the "Modified Swedish Classification" (Table 1.3.2) :

Table 1.3.2 Modified Swedish Classification

Spastic	Hemiplegia
	Tetraplegia
	Diplegia
Ataxic Diplegia	
	Congenital (simple)
Dyskinetic	mainly choreoathetotic
	mainly dystonic

after Mutch et al 1992

As will be seen later, even this simplified classification contains inherent pitfalls for the clinician in relation to selection for treatment, particularly with regard to the understanding of the pathophysiology of "spasticity" which is still considered by most authors and paediatricians to represent a motor syndrome rather than a specific measurable, pathophysiological entity which can be operationally defined. Crothers and Paine (1959, p 37) adapted the classification of Wagley (1945) in their definition of "spasticity" which is far more inclusive than the physiological operational definition of spasticity as a velocity-dependent increase in stretch reflexes commonly used in adult neurology (Lance, 1980). According to Crothers and Paine (1959), "spasticity" is defined as a syndrome comprising:

1 Muscular hypertonus of the clasp-knife type
2. Hyperreflexia, which is reproducible on repeated elicitation
3. Positive Babinski and Hoffman reflexes
4. Diminished superficial reflexes (abdominal and cremasteric)
5. Release of postural and labyrinthine reflexes
6 Spread and overflow of associated movements
7. Loss of voluntary control of fine finger movements
8. Frequently clonus of ankle or other joints (although not universal or essential)
9. Tendency to muscular contracture in characteristic postures.

after Crothers and Paine 1959, p37.

Accordingly, this definition of *syndromic spasticity* is a mixture of observations relating to reflexes, postures, selective motor control, involuntary movements and natural history. Some of the elements of this definition of spasticity can therefore only be applied *a posteriori*, ie retrospectively. The inclusion of postural and labyrinthine reflexes in the definition of "spasticity" commonly used for children with cerebral palsy (see also Denny-Brown, 1980) may be one of the reasons for the disappointing results from the use of specific 'antispastic treatments' (see section 1.5: treatments in CP).

A further confusion has been the equation of spasticity with the anatomical loss or disruption of the "pyramidal" fibres or tracts, an evident misnomer since lesions of the medullary pyramid most likely result in hypotonia, hyporeflexia, loss of distal, fine, voluntary motor control (Towers, 1940; Bucy,1964 see also Brodal, 1981, p182-194) and loss of 'cortical reflexes' such as the positive supporting, placing and hopping reactions (Rademaker 1931, quoted by Crothers and Paine, 1959). Accordingly, cerebral palsy phenotypes quoted in the literature for the purposes of epidemiological or clinical classification include a variety of

aetiological, anatomical, physiological, developmental and natural historical assumptions regarding definition, cause(s) and prognosis of cerebral palsy. As discussed in the foregoing sections, this renders difficult the task of case -definition and prospective follow-up, especially in the context of specific treatments with putatively specific mechanisms of action. Hagberg and Hagberg (1993) have used the "Swedish Classification" of cerebral palsy to produce a breakdown of the frequency of topographical CP syndromes (Table 1.3.3):

Table 1.3.3 The West Swedish CP Series Birth Years 1979-1986: Gestational Age (GA) Groups related to Syndromes.

GA	<32		32-36		<37		>37		Total	
Syndrome	n	%	n	%	n	%	n	%	n	%
Hemiplegia	5	13	15	22	24	18	88	46	112	34
Diplegia	51	74	42	62	93	68	51	27	144	44
Dyskinetic	1	1	6	9	7	5	21	11	28	9
Tetraplegia	5	7	4	6	9	7	14	7	23	7
Simple Ataxia	3	4	1	1	4	3	17	9	21	6
Total	69	100	68	100	137	100	191	100	328	100

after Hagberg and Hagberg 1993.

The distributions of the CP phenotypes quite clearly relate to gestational age. For the term infant (GA>37 weeks), hemiplegia is the dominant syndrome affecting just under half of all cases followed by diplegia with just under one third of cases leaving dyskinesia, tetraplegia and simple ataxia to account for less than 10% of cases respectively. Under 37 weeks GA, diplegia replaces hemiplegia as the dominant motor syndrome accounting for 68% of cases and the dyskinesias and simple ataxias each represent less than 5% of the CP population respectively. Taken as a whole, irrespective of gestational age, diplegia is the commonest CP phenotype accounting for 44% of all cases, followed by hemiplegia with 34%, dyskinesia at 9%, and tetraplegia at7% trailing as joint equal with simple ataxia at 6% of all cases. The bilateral CP syndromes account for 54% of cases born at 37 weeks and over, rising to 88% of cases under 37 weeks gestation, diplegia becoming the dominant phenotype with diminishing gestational age.

Another example of the application of a topographical classification scheme is given in table 1.3.4 which has been used to classify the neuromotor impairment of children surviving grade III or grade IV periventricular brain haemorrhage.

Table 1.3.4 Topographical distribution of neurological signs and pattern of clinical tone abnormality following grade III or IV intraventricular haemorrhage

Topography	Normal	Dominant Pattern of Clinical "Tone" Spastic	Dystonic	Ataxic hypo-tonic	Mixed	Total
Hemiparesis						
R	5	0	0	0	1	6
L	2	0	0	0	0	2
Diplegia	0	0	3	2	1	6
Triplegia						
R	0	0	2	1	0	3
L	0	0	2	0	1	3
Quadriplegia	0	1	1	1	0	3
Total	7	1	8	4	3	23

after Lin et al 1993.

The salient neurological findings were that 10/33 appeared neurologically intact; asymmetrical injury (grade IV IVH) did not carry a worse prognosis than symmetrical injury (grade III IVH); the commonest neurological phenotype was hemiplegia in 8/33 followed by diplegia in 6/33, triplegia in 6/33 and total body involvement in 3/33 cases. As might be expected, hemiplegia and triplegia were associated with a grade IV IVH (with extension of haemorrhage into brain parenchyma) whereas diplegia and tetraplegia occurred in cases with symmetrical ventriculomegaly: ie periventricular leucomalacia or white matter loss (see below) A striking feature of these studies was that the pattern of muscle tone was normal in 7/8 of the hemiplegic children, none of whom exhibited pure clinical "spasticity" although 1/8 cases showed mixed signs. None of the diplegic children were considered "spastic" but showed evidence of "dystonia" (3/6), ataxia / hypotonia (2/6) and a mixed picture in 1/6 cases. A similar distribution was seen in the six triplegic children, though the 3 quadriplegic cases spanned the spastic, dystonic and ataxic / hypotonic groups as single cases.

One inherent misnomer enshrined in the current definition of cerebral palsy is the *a priori* exclusion of disorders of other parts of the nervous system, clearly refuted by the following illustrative example (fig. 1.3.1), which shows a well defined left hemisphere porencephalic cyst secondary to cerebral infarction (top, white arrow) sometime after 22 weeks gestation, since gliosis is present. In contrast with the opposite side, the internal capsule (top, thin black arrow) of the damaged hemisphere is thin and atrophic and the contralateral medullary pyramid (bottom, arrow) is virtually absent indicating that the

destruction of higher structures is likely to cause anatomical rearrangements lower down in the central nervous system: in this case an almost total loss of descending corticospinal fibres. This issue of damage or transformation of the motor system "downstream" of the lesion, and of central and peripheral interactions and adaptations is a recurrent theme of the present work.

The relationship between damage in the brain and possible damage in the spinal cord has also been critically examined by Crawford and Hobbs (1994) on the basis of careful correlation of the reported pathological changes in periventricular leucomalacia (PVL) and current magnetic resonance imaging (MRI) techniques. Hobbs and Crawford (1994) draw attention to the lack of evidence for "anoxic" damage as the mechanism of diplegia of prematurity (although Polani, 1958, was concerned with the possible association between hyperoxia and both diplegia and the retrolental fibroplasia in preterm infants exposed to high oxygen regimens). These authors point to the fact that the principal sites of white matter wasting as seen in pathological specimens and on MRI scans occur at sites remote from the corticospinal tracts running in the posterior limb of the internal capsule: indeed the sites classically associated with the PVL of prematurity are in the region of the interventricular foramen (anterior to the corticospinal tracts) and the collateral trigone and posterior horn of the lateral ventricles (some distance posterior to the corticospinal tract). What Hobbs and Crawford propose instead, is a dying back hypothesis of the motorneurone axons with a predilection for the longest axons, ie those supplying the legs. Whether such a process would affect the cell bodies in the brain or begin in the descending fibres themselves is not known. Whatever the eventual mechanism(s) of PVL, changes of one sort or another in the spinal cord are likely.

The correlation between observable brain damage on neuroimaging (CT scanning) and functional impairment is not necessarily tight. Indeed one of the striking features of an extensive Swedish clinical and radiological study in 111 cases of childhood hemiplegia by Wiklund and Uvebrandt (1991) indicated the lack of a tight correlation between specific cerebral tissue volume loss and the loss of a wide range of motor and non-motor functions although these authors found statistically significant correlations between mild clinical dysfunction and the 29 normal CT brain scans. Cerebral maldevelopment (n=19) correlated weakly with establishing the dominant limb ($p< 0.03$) and growth impairment in the leg ($p<$

0.017) while for the 47 cases with periventricular lesions, the only significant correlation was with growth impairment of the arm ($p< 0.015$). Only 13 cases exhibited cortical / subcortical damage which, somewhat surprisingly, did not correlate with gait, growth, intellectual, speech, hearing or visual impairments. This type of injury, did however correlate with impaired hand function ($p<0.002$), general motor function ($p< 0.03$), stereognosis ($p<0.001$), two-point discrimination ($p<0.001$), facial weakness ($p< 0.001$) and epilepsy ($p<0.024$).

Another interesting finding was that 13 cases (12%) had evidence of bilateral lesions. Such studies demonstrate the heterogeneity of the the clinical phenotypes collectively referred to as childhood hemiplegia, indicating the need for caution in generalising between cases. This preservation of function in the presence of conspicuous brain damage has been described by Lin *et al*, 1993, following grade III and IV intraventricular / periventricular haemorrhage and is illustrated in fig1.3.2. Severe CP may be present with normal imaging and function preserved despite conspicuous loss of brain substance.By contrast, small lesions may be profoundly disabling as shown in fig. 1.3.3, which illustrates a case of hemidystonia in a 12 year old girl with a congenital infarct of the right globus pallidus.

Associated complications include epilepsy, which accompanies one third of all cases: 50% in the case of hemiplegic CP and even higher in quadriplegic CP. The risk of subsequent learning difficulties is much higher if CP is complicated by epilepsy and conversely, the prognosis for educational development is far higher in the absence of fits. A lowering of the IQ of children with hemiplegia was strongly correlated to a history of current or resolved seizures, acquisition early between 1 and 60 months of life (as opposed to "congenital" or acquisition after 60 months of life), bilaterality, severity and head circumference which all acted as independent predictors of IQ (Goodman and Yude 1996). As a distinct group, cases with pure basal ganglia (extrapyramidal) syndromes, though severely motor impaired may nevertheless have normal intelligence.

<u>A note on kernicterus: a model for prevention.</u>

The association between neonatal jaundice and damage to the basal ganglia has been clearly understood since Schmorl's original description of "kernicterus" in 1904 causing:

..."damage to the subthalamic nuclei, Ammon's horn, globus pallidus, inferior olive, cranial nerve nucleii in the floor of the 4th ventricle and in the dentate nucleii, flocculi and cerebellar vermis".

Christensen and Melchior 1967 p 9

Later experiments in monkeys by Lucey and colleagues (1964) produced nuclear jaundice only when associated with asphyxia. A clear difference between the cerebral palsies associated with perinatal jaundice, is its association with the characteristic "ski slope" hearing deficit in which a high frequency loss is typical of sensorineural deafness (Crothers and. Paine, 1959, p 139.).

A final comment on the pathogenesis of kernicterus is that with the advent of the Rhesus isoimmunisation programme, Rhesus incompatibility as a cause of severe neonatal jaundice in the second child born to rhesus incompatible parents has all but disappeared, as have the majority of cases due to ABO incompatibility. This is probably the only single type of cerebral palsy which has been actively prevented by modern obstetric practices, although haemolytic jaundice due to other causes is still a predisposing factor.

Non-progressive basal ganglia damage producing severe dyskinetic cerebral palsy with preservation of cognitive abilities is a well recognised clinical entity among term infants. Although affecting only about 9% (10/115) of all cases (Rosenbloom 1994), this form is attributable to a late, sudden, severe intrapartum hypoxic-ischaemic insult together with an immediate severe depression and acidosis requiring resuscitation but resulting in only a mild to moderate neonatal hypoxic-ischaemic encephalopathy (see Sarnat and Sarnat 1976) with recovery within 7 days, indicating a relative sparing of cortical structures. Overall, Rosenbloom (1994) found a total of 115/1056 cases (ie 11%) of dyskinetic CP on the Merseyside (UK) Cerebral Palsy Register, so this particular group form only 1% of the total population. The pathology of the basal ganglia disorders is reviewed by Christensen and Melchior (1967, p 7-9 and 97-107) and the radiology of three cases is reviewed by Rutherford and colleagues (1992). Unlike the reports in the survey of the literature by Christensen and Melchior in which none of the cases of CP with basal ganglia lesions were cystic, Rutherford *et al* (1992) report three case of haemorrhagic basal ganglia lesions attributable to HIE. These babies were initially hypotonic in the neonatal period, becoming hypertonic with jittery and tremulous movements soon after withdrawal of anticonvulsants. The authors comment that this is the reverse of the classical HIE picture of prolonged hypotonia with persistence of primitive reflexes, or of the clinical picture in kernicterus in which athetoid movements emerge after the first year of life.

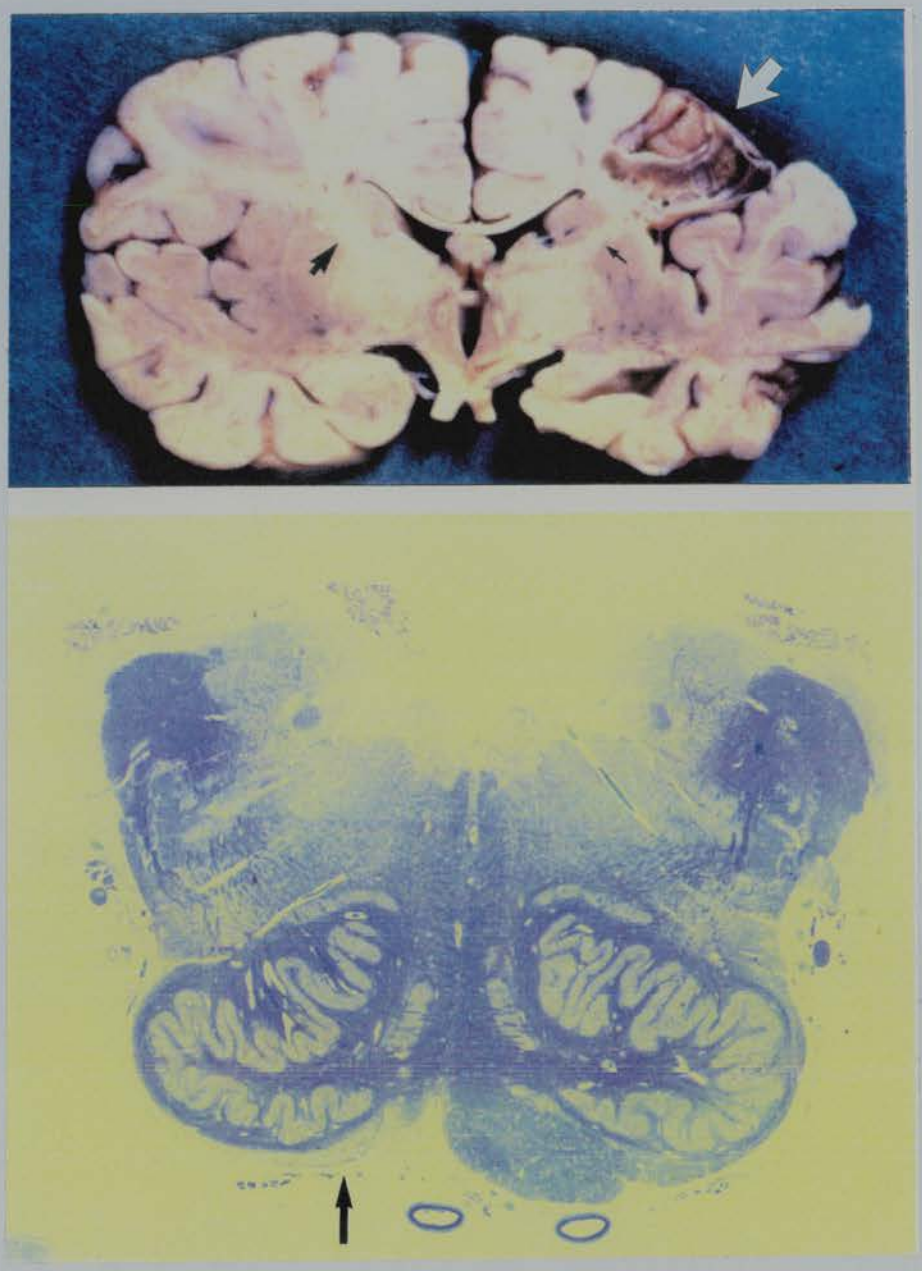


Fig. 1.3.1. Anatomical correlations of cortical-subcortical infarction.
In this four month old infant brain, the internal capsule (thin black arrow) of the damaged hemisphere is thin and atrophic compared with that of the opposite hemisphere and the medullary pyramid contralateral to the damaged hemisphere (thick black arrow) is virtually absent, indicating that the destruction of higher structures is likely to cause anatomical rearrangements lower down in the central nervous system such as the spinal cord.
By permission of Dr JK Brown, Royal Hospital for Sick Children, Edinburgh.

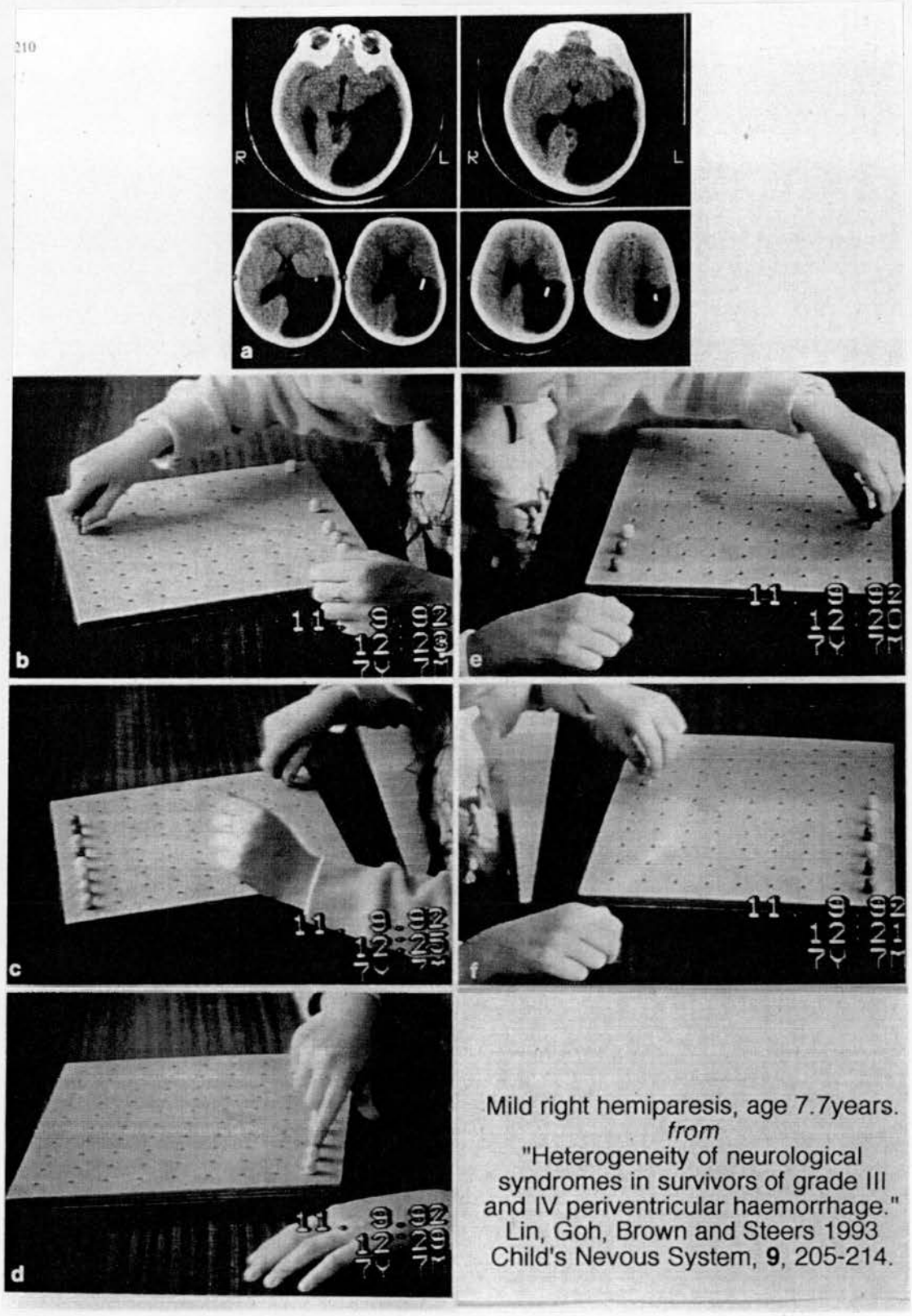


Fig 1.3.2. Lack of clear correlation between extent of brain volume loss and function.
Peg board testing of the hemiparetic right hand is still possible despite gross destruction of the opposite left hemisphere in an ex-preterm infant surviving grade IV *by permission.*

Fig 1.3.3 Discreet congenital infarction of the right globus pallidus.
The lesion is shown with a white arrow as an area of high signal intensity on an MRI scan using T2-weighted images. The lesion has the same signal intensity as the cerebrospinal fluid. This tiny lesion gives rise to a congenital left hemidystonia: case 23, see sections 3, 5 and 6.

1.4 Natural History of Cerebral Palsy

The inherent developmental potential for improvement in motor function in children with cerebral palsy is the chief factor acting favourably on the child's behalf, outstripping any likely benefit from the large numbers of treatments available today. The rate of this development, it's extent, quality or liability to arrest or "reverse" is referred to as the "natural history" of the condition, which results from complex changes in central nervous and musculoskeletal system mediated by the processes of growth, maturation, healing and the emergence of compensatory strategies for motor control in the wake of cerebral damage.

With the exception of walking, little is known about the natural history of cerebral palsy. Crothers and Paine (1959) were among the first to chart the prognosis for walking according to topographical phenotypes and characterisation of the type of predominant motor dysfunction (fig. 1.4.1). According to their data, 75% of hemiplegic children walk by the age of 2 years, compared with only 25% of children exhibiting extrapyramidal or mixed CP motor patterns or those with tetraplegia. By four years, half of the extrapyramidal/ mixed group of children are walking compared with 95% of hemiplegic children. By the age of 14 years, all hemiplegics are walking compared to 80% of the extrapyramidal / mixed group and 70% of the tetraplegic children.

The factors relating to the walking prognosis have been studied and reviewed by Crothers and Paine (1959), Beals (1966), Bobath (1966), Bleck (1975), Molnar and Gordon (1976), Badell-Ribera (1985), Watt *et al* (1989), Campos da Paz et al (1994) and Trahan and Marcoux (1994) . These studies have been reviewed by Sala and Grant (1995)., the principal favourable and adverse prognostic factors for walking falling into three main groups: 1. The retention of primitive reflexes and absence of postural reactions are associated with a poor ambulatory potential (table 1.4.1). 2. The acquisition of certain gross motor skills such as sitting by the age of 2 years or reciprocal crawling by the age of 30 months is favourably associated with independent community ambulation (table 1.4.2). 3. The type of cerebral palsy, including topographical distribution and dominant pattern of tone influence the potential for ambulation (table 1.4.3).

These studies indicate that the prognosis for ambulation is adversely affected by the retention of primitive reflexes (asymmetric and symmetric tonic neck, moro, neck righting, tonic labyrinthine, extensor thrust and positive supporting reflexes) and the absence of

postural reactions such as the foot-placing and parachute responses. Positive prognostic indicators being the attainment of unsupported sitting between 1-2 years of age and reciprocal crawling by 30 months of age.

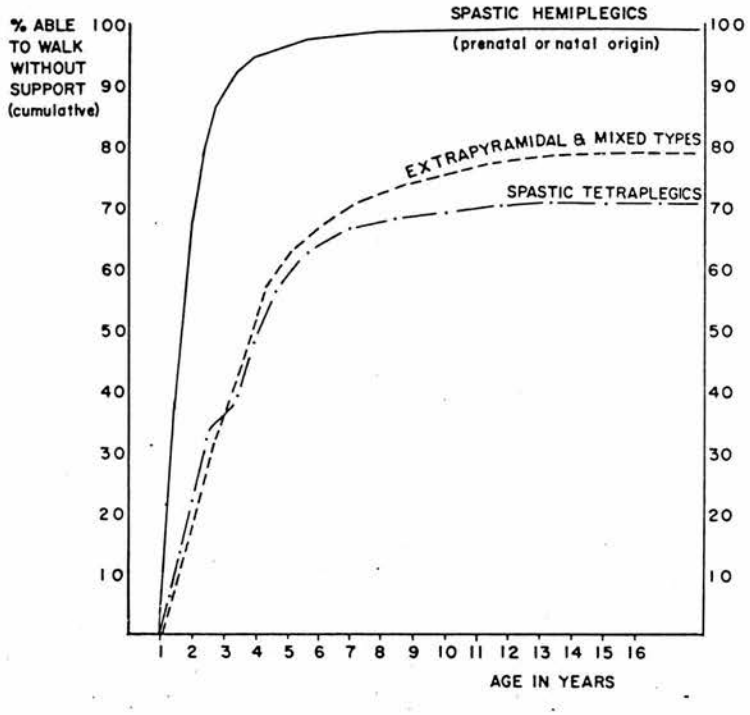


Fig. 1.4.1 Age of walking without support of cerebral palsy patients (n=289).

Crothers and Paine 1959 with permission, p.119.

The cerebral palsy phenotype also indicates the likely attainment of walking, which is achieved in 100% of spastic hemiplegic (Crothers and Paine, 1959; Bleck, 1975; Molnar and Gordon, 1976 and Watt *et al*, 1989) and ataxic children (Molnar and Gordon, 1976 and Watt *et al* ,1989), 90% of spastic diplegic (Beals 1966; Molnar and Gordon, 1976; Badell-Ribera, 1985; Watt *et al*, 1989; Campos da Paz *et al*, 1994 and Trahan and Marcoux, 1994) and 87% of spastic triplegic (Campos da Paz *et al*, 1994) 72% of spastic quadriplegic children (Molnar and Gordon, 1976; and Watt *et al*, 1989; Campos da Paz *et al*, 1994) and 71 % of children with athetosis (Molnar and Gordon, 1976; Watt *et al*, 1989). This compares with only a 50% chance of ambulation in mixed spastic-athetoid CP (Molnar and Gordon, 1976) or non-ambulation in hypotonic CP (Molnar and Gordon 1976; and Watt *et al*, 1989).

Table 1.4.1 Persistent primitive reflexes and absent postural reactions associated with non-ambulatory status.

Clinical signs examined		
Primitive Reflexes	*Association with inability to ambulate*	
Asymmetrical Tonic Neck Reflex	1 of 5 most frequent signs	(Bleck 1975)
	Associated	(Crothers 1959)
	If present at 12, 18 and 24 months	(Molnar 1976)
	Significant association	(Watt et al 1989)
Symmetrical Tonic Neck Reflex	If present at 12, 18 and 24 months	(Molnar 1976)
	Associated	(Trahan 1994)
	Significant association	(Watt 1989)
Moro	1 out of 5 most frequent signs	(Bleck 1975)
	If present at 12, 18 and 24 months	(Molnar 1976)
	1 of 3 most strongly associated	(Trahan 1994)
	Significant association	(Watt 1989)
Neck Righting	1 out of 5 most frequent signs	(Bleck 1975)
	No significant association	(Watt 1989)
Tonic Labyrinthine	If present at 12, 18 and 24 months	(Molnar 1976)
	1 of 3 most strongly associated	(Trahan 1994)
	Significant association	(Watt 1989)
Extensor Thrust	1 out of 5 most frequent signs	(Bleck 1975)
	No significant association	(Watt 1989)
Positive Supporting	If present at 12, 18 and 24 months	(Molnar 1976)
	Associated	(Trahan 1994)
Postural Reaction	Association with inability to walk	
Absent Foot Placement	Significant association	(Watt 1989)
Absent Parachute Reaction	1 out of 5 most frequent signs	(Bleck 1975)
	Significant association	(Watt 1989)

after Sala and Grant 1995.

Table 1.4.2 Age of attainment of gross motor skills quality of walking

Achieved gross motor skill	Functional Level of Ambulatory Status	Authors
Arm propping to sit at 1.5-2yrs	Exercise only ie requiring assistance to stand and assistive devices to ambulate.	
Arm propping <1.5 yrs Sat from prone by 2 yrs Crawled symmetrically by 2.5 yrs	Household / low endurance	
Sat from prone and Crawled symmetrically at 1.5-2.5 yrs Crawled reciprocally after 3 yrs	Household / high endurance	
crawled reciprocally at 1-2.5 yrs	Community ambulators	Badell-Ribera n=50, (1985)
Head balance in prone by 9 months (but not>20 months)	Independent or with assistive device	
Sitting by 24 months (but not after 36 mths)	Independent or with assistive device	
Crawling Symmetrically or asymmetrically by 30 mths (but not after 61 mths)	Independent or with assistive device	Campos da Paz n=272, (1994)
Sitting by 2 years	100% community ambulators	
sitting by 2-3 years	55% community ambulators	
sitting after 3 years	Household ambulators only	Molnar and Gordon n=164, (1976)
Sitting by 2 years	Extrapyramidal CP: 97% ambulant Spastic tetraplegia CP: 90% ambulant	
Sitting by 2-4 years	Extrapyramidal or spastic tetraplegic CP Only 50% ambulant.	Crothers and Paine n=138, (1959)

after Sala and Grant 1995

Table 1.4.3 Type of cerebral palsy and the attainment of walking.

Type of CP	% ambulant	n	Authors	
Spastic Diplegia	91	108	Campos da Paz *et al*	(1994)
	90	50	Badell-Ribera	(1985)
		83	Beals	(1966)
		31	Watt et al	(1989)
	87	115	Trahan & Marcoux	(1994)
	86	37	Molnar and Gordon	(1976)
Spastic Quadriplegia	70		Crothers and Paine	(1959)
	72	85	Molnar and Gordon	(1976)
	27	37	Campos da Paz *et al*	(1994)
	0	21	Watt *et al*	(1989)
Spastic triplegia	87	13	Campos da Paz *et al*	(1994)
Spasticity-athetosis	50	28	Molnar and Gordon	(1976)
Athetosis	71	14	Molnar and Gordon	(1976)
	0	2	Watt *et al*	(1989)
Extrapyramidal	80		Crothers and Paine	(1959)
Ataxia	100	6	Molnar and Gordon	(1976)
		1	Watt *et al*	(1989)
Hypotonia	0	2	Molnar and Gordon	(1976)
		1	Watt *et al*	(1989)

after Sala and Grant 1995.

As far as can be ascertained, there is no evidence for any change in the natural history of recognised CP syndromes over the last 40 years. What is apparent, is that the case mix comprising the CP population has changed (ie decline of kernicterus and rise in the white matter disease (s) of prematurity/ low birthweight). Section (1.5) deals with the issue of specific treatments in cerebral palsy.

1.5 Treatments in Cerebral Palsy

One measure of the success of treatments in cerebral palsy would be evidence of a change in the natural history of the motor manifestations in the same way as attempts to find evidence that changes in obstetric practices "prevent" cerebral palsy. It may be pertinent to ask if the natural history of cerebral palsy has changed since Crothers and Paine wrote "*The Natural History of Cerebral Palsy*" in 1959. In their book, the authors are anxious to indicate the mismatch between parental expectation, the medical zeal or appetite for intervention, the innate potential for ambulation and therapeutic outcome. Despite the almost 40 years which

have elapsed since Crothers and Paine wrote their book, there is no evidence of any treatment, either singly or combined, altering the natural history of cerebral palsy, partly owing to the methodological pitfalls inherent in the field, chief amongst which is the clinical heterogeneity of the cases (see also physiological studies by Brown et al (1987, 1991) and Lin and Brown (1992):

> "The only way to come to any conclusion is to try to find groups of fairly comparable cases, consider the information we have about the usual course of growth and development and the influence of the various types of management upon them. In this way it is possible, by various highly vulnerable analogies, to speculate more profitably on the problems of the confused groups which are not composed of comparable individuals."
>
> *Crothers and Paine, 1959, p174.*

1.5.1 Aims of treatment.

The overall objective of treatment in cerebral palsy (CP) is to improve function and prevent deformity (Brown and minns, 1989). As seen above, the phenomena which clinician are attempting to alter are extremely complex. The successful outcome of any intervention depends on a number of factors including:

1. Case definition.
2. Understanding the pathophysiology of the underlying problem.
3. Understanding the mechanism of action of the proposed treatment.
4. Objective (operationally defined) methods of evaluation.
5. Criteria for the success or failure of the treatment (treatment goals).

A comprehensive review of treatments in CP (Table 1.3.5.1) testifies to fundamental gaps in our understanding of the pathophysiology of CP

(see Lin, Brown and Brotherstone 1994 a, 1994 b).

Table 1.5.1. Treatments in Cerebral Palsy.

1. Physical Therapies	(Bobath, Voitja, Domen-Delicato, Peto, high resistance exercise).
2. Orthotics.	(solid and hinged, floor reaction, nocturnal)
3. Plaster Immobilisation.	
4. Drugs	(diazepam, baclofen:oral and intrathecal, dantrolene, L-dopa, nerve blocks).
5. Botulinum Toxin.	(intramuscular injections)
6. Therapeutic Electrical Stimulation.	
7. Functional Electrical stimulation	(Timed with movements)
8. Orthopaedic surgery	(single and multiple soft tissue releases with or without bone surgery).
9. Selective dorsal rhizotomy.(SDR)	(multiple divisions of dorsal sensory afferent rootlets)

The use of physical therapies combined with orthotics with occasional plaster immobilisation and surgery remain the pillars of interventional management in CP, although

much remains to be learned regarding the effects of such treatments on the muscles and the nervous system. Muscle strengthening as an avowed aim of treatment has still to enter the framework of routine care in contrast to "tone lowering" by slow passive stretching. Whatever the overall approach, it must be borne in mind that muscles are designed to work and to move, and one aim of treatments should be to promote movement. Scrutton (1984) has brought together some of the ideas underlying the physical therapies and the idea of aim-oriented approaches; Bleck (1987; 1990) offers some practical and critical reviews on current surgical practices. These works have been supplemented by multi-author texts hoping to bring together current thinking in motor physiology with treatment practices (Forssberg and Hirschfeld, 1992; Sussman, 1992).

The three treatments which have received most attention in the past ten to fifteen years are intrathecal Baclofen, selective dorsal rhizotomy and intramuscular botulinum toxin A injections. These will be briefly examined in turn with regard to:

1. site of action
2. safety
3. efficacy
4. complications
5. practicality
6. repeatability
7. cost.

1.5.2.Intrathecal Baclofen.

i. The basis of action of baclofen.

Baclofen is a GABA-b receptor agonist which was first used in the treatment of spasticity in 1970 at the Prince Henry Hospital in Australia in two quadriparetic and 4 quadriplegic adults over a 3 month period. The study concluded that baclofen reduced the velocity sensitivity to stretch (dynamic sensitivity), reducing muscle spasms in 3/6 and clonus in 2/4 cases without causing weakness (Jones *et al* ,1970). Overall, baclofen proved superior to diazepam (a GABA- a receptor agonist) in 5/6 cases. A further open trial in 115 patients with spinal and cerebral lesions (Jones and Lance, 1976) using larger doses for up to six years, demonstrated that baclofen was largely beneficial to those patients with spinal spasticity in whom 87% (69/79) had improved and in 52% (41/79) spasticity was "no longer a problem" so that the limiting factors to mobility were the underlying weakness produced by the paraparesis or paraplegia.

Complications included: dreams, hallucinations or visual illusions (8/110), drowsiness (8/110), depression (2/110), paranoia (2/110), headache (2/110), blurred vision (2/110) nausea (2/110) tremor (2/110): ie cumulative side effects were noted in 20% of cases receiving treatment. There were no biochemical or haematological side effects, but one case attempted suicide by taking 1000 mg of baclofen which produced coma requiring ventilation for six hours associated with hypotonia, areflexia and flexor plantar responses. All the features of spasticity returned as the baclofen wore off.

This early trial demonstrated potential benefits in the use of baclofen but already indicated its limited usefulness in cerebral motor disorders, potential "central" side effects and the risks of over-dosage leading to respiratory depression. Further studies confirmed that the GABA-b receptors are concentrated in the presynaptic endings of afferent fibres entering the dorsal horns of the spinal cord (Price *et al*, 1984, 1987; Bowery, 1989) which accords with the view that spasticity arises in part as a result of a loss of presynaptic inhibition (Burke and Ashby, 1972).

Burke, Andrews and Gillies (1971) were able to demonstrate an increase in the reflex velocity threshold of the quadriceps and hamstrings muscles with passive sinusoidal stretch (see section 5, below).

Another interesting functional study of 11 spastic patients was reported by McLellan (1977) who were selected for co-contraction patterns seen with surface EMG recordings from the quadriceps and hamstrings muscles during voluntary flexion and extension movements at the knee. Most patients with brisk tendon reflexes and increased muscle tone, extensor plantar responses, only mild gait gait impairment and relatively well-preserved strong voluntary power showed little evidence of co-contraction during this task. The rates of voluntary flexion and extension at the knee seldom exceeded 1Hz (being 3-4hz in healthy control adults): these rates were also slower than the threshold rates required to elicit a stretch-reflex (see section 5, below). Out of 32 subjects screened, 14 exhibited co-contraction during the above task and in whom also passive stretches at the same rate provoked a stretch reflex. Of these 14, 11 had usable recordings and the subjects performed sinusoidal tracking tasks, 10°-80° from full knee extension, before and for 4 hours after 20mg of oral baclofen. McLellan found that during voluntary movements, in mild to moderate cases, the stretch reflex was suppressed whereas in severe spasticity, the stretch reflex was

enhanced. The reflex response to passive stretches was reduced by 30% when plasma levels reached 250ng/ml and to over 50% at concentrations of 400ng/ml:however, this effect was completely overridden during voluntary movements when co-contractions reappeared, indicating that the mechanisms of co-contractions are different to those of spasticity and passively elicited stretch reflexes.

ii. Trials of intrathecal baclofen: patient selection and complications.

Intrathecal infusions (IT) of baclofen emerged as a strategy for overcoming the central effects produced by large doses and chronic use. The first report showed improvements in the Ashworth Scale (1964) in 6 adults who had programmable pumps implanted (Penn and Kroin, 1985). Technical difficulties included problems of pump implantation, programming, over-dosage causing lightheadedness, weakness, drowsiness and loss of consciousness requiring ventilation. Catheter blocks and leaks were also reported. Subsequently , seven adult patients were maintained on IT baclofen infusions for up to 2 years, during which, muscle tone was reduced to normal and muscle spasms ceased with an improvement in sleeping pattern. Benefits included abolition of spasms, which allowed cases to ambulate with long-leg braces or crutches in 2/7 cases and improved bladder function in 3/7 cases, resolution of urinary incontinence in 4 cases and 3 cases returned to work: IT baclofen doses varied from 15-650µg/day. Rehabilitation that had been abandoned in 3 cases because of rigidity, was restarted.

Complications included: revisions to pump pocket site, faulty pump in 2 cases, and drug overdose in 2 cases receiving bolus dose administrations of IT baclofen, in which patients experienced lightheadedness, upper limb weakness and coma over a 30-60 minute period. The cerebrospinal fluid (CSF) concentrations of baclofen reached 4.8 and 11.3 µg/ml, with a half-life of 5 hrs. As the effects of the baclofen wore off, the legs experienced violent spasms, while remaining hypotonic between spasms. These spasms settled as the background tone returned over three days. The patients resumed infusions by continuous mode thereafter.

Overall the four spinal cord-injured patients experienced the greatest benefit, whereas the three multiple sclerosis patients, with more upper limb impairments, fared less well: in no case was voluntary function restored to any great degree.

This study was then followed up by a double-blind cross-over study of IT baclofen

against IT saline infusions for three days in 20 patients with severe spinal spasticity (Penn *et al*, 1989) during which neurological examinations , motor function assessments and patient assessments correctly identified periods of baclofen infusion: all patients then entered a long-term open trial of IT baclofen infusions, and in all cases, tone, as assessed by the Ashworth scale was reduced.

Another report studying the H-reflex in six adults with post-traumatic spinal lesions undergoing IT Baclofen infusions, demonstrated a reduction in the Hmax/Mmax ratio in 4/6 cases (Macdonell et al, 1989) along with reductions in the Ashworth scale.

The benefits of IT baclofen for patients with spinal injury as opposed to multiple sclerosis was reported by Lazorthes and colleagues (1990) who evaluated 38 cases with bolus injections administered via a port and proceeded to chronic infusions in 18/38 of these. They reported significant functional improvements in 9/18 cases which was considerable in three. The selection for chronic infusions is shown in table 1.5.2:

Table 1.5.2 Selection of cases for chronic IT baclofen infusions.

Aetiology	IT bolus Trial	Chronic Administration
Spinal spasticity		
multiple sclerosis	12	6
degenerative myelopathy	2	0
spinal trauma	14	7
spinal ischaemia	1	1
transverse myelitis	1	1
Cerebral spasticity		
cerebral palsy	5	1
brain trauma	2	1
ischaemic lesion	1	1
Total cases	38	18

after Lazorthes et al, 1990

Once again, Lazorthes and colleagues found the best improvements in cases with incomplete, post-traumatic spinal paraplegia, the multiple sclerosis patients doing less well: the single case of cerebral palsy proceeding to chronic infusions of IT baclofen experienced a moderate improvement, though this was curtailed by pump failure and over dosage resulting in the treatment being stopped. Overall , overdose occurred in 2 cases, catheter displacement in 3, meningitis in 3 and sepsis in 2 cases respectively. Treatment was stopped in 9/18 cases because of one or more of these complications, although in one case, no

complications were reported. It can be seen that the pre-selection of patients with a period of bolus injections is costly in time and resources, and only about half of cases proceeded to chronic infusions which were discontinued in a further half owing to complications.

iii. IT baclofen and the movement deficit.

The studies of McLellan (1977) on the effects of oral baclofen on co-contractions during voluntary alternating movements at the knee have been mentioned above: he found that whereas the passively elicited velocity-dependent stretch reflexes were reduced with oral baclofen, co-contractions with voluntary movements was unaffected.

Latash and colleagues(1989) studied six cases undergoing IT baclofen with long-standing spasticity associated with spinal cord pathologies and concluded that the tonic co-activation of muscles was decreased whereas voluntary agonist contractions remained unaffected by the infusions, one case being able to perform fast, isotonic,alternating movements: this study was taken as part proof that useful voluntary function could be 'unmasked' once the effects of disabling spasticity was controlled. Nevertheless, the McLellan and Latash studies are conflicting.

iv. IT baclofen in children

As is evident from the preceding reports, IT baclofen appeared most useful in spinal pathologies and was most effective in cases of spinal trauma as opposed to demyelinating diseases: overall cases with 'cerebral spasticity' fared less well. Albright and colleagues have applied the principle of single bolus injections of IT baclofen in children with cerebral palsy.

A dose-ranging, single dose, double-blinded IT baclofen study involving 17 cases (aged 5-31 years) with congenital, moderate to severe spastic quadriplegia (Albright *et al*, 1991) showed falls in the Ashworth scale within two hours of IT bolus injections although the ordinal Ashworth scale was handled as if it were a continuous parametric scale. Boluses of 25μg caused no unwanted side effects. In 2/17 of the youngest and lightest children, the 50μg boluses caused self-limiting lethargy in 1 case, 4-6 hours post-injection and disorientation, agitation and lethargy, 9 hours after injection in another. Upper extremity tone and function were not significantly affected by single dose bolus injections. However, there is no mention of the functional benefit derived from the bolus IT baclofen: the authors concluded that single dose bolus studies might help identify potential candidates for selective dorsal rhizotomy!

Chronic trials of IT baclofen have been reported by Armstrong and colleagues (1992) in two ventilator-dependent children with chronic post-traumatic mixed cranial and spinal spasticity, spasms and rigidity. Both children had been given trials of oral baclofen but developed symptoms of sedation at attempted increases in the dose (30mg/day for one case and 100mg/day in the other case). The first case showed evidence of gradual tolerance to IT baclofen which culminated in doubling the daily IT dose from 600µg/day to 1200µg/day over a 16 month period. Problems with breakthrough spasms resulted in myelography which demonstrated a spinal block to flow from an arachnoiditis attributed to the original injury 6 years previously. Breakthrough spasms eventually resulted in a selective dorsal rhizotomy at 18 months into IT baclofen therapy. One year after rhizotomy, the upper limb dystonic posturing returned, but was again controlled by IT baclofen. The second case died following a bolus infusion of 800µg of IT baclofen after a partially beneficial trial of IT baclofen which had necessitated may revisions of the infusion pump concentration and infusion rate over an 18 month period. A post mortem in the second case revealed inflammatory changes in the lumbar spinal meninges and in the posterior horns of the spinal cord indicating long standing inflammation. (see also Armstrong, 1992, for an early review)

v. Summing up of IT baclofen in cerebral palsy.

Both of the above cases by Armstrong and colleagues (1992) illustrate the complexity surrounding continuous IT baclofen infusions along with the associated morbidity and possible mortality arising from over-dosage and infection. The Albright series (1990) suggests that IT baclofen has a place in selecting children who might benefit from rhizotomy as well as chronic infusions.

Overall the costs and technical difficulties of IT baclofen infusions, cast doubts on the advisability of IT baclofen therapy in cerebral palsy without a full evaluation service: in specialist hands it may have a place in severe spinal spasticity. However, the years of baclofen evaluation have shed light on the pathophysiology of spasticity and spurred research into GABA receptor type, distribution and function.

1.5.3. Selective dorsal rhizotomy (SDR).

Peacock and Staudt (1990) have strongly advocated the use of SDR whereas its place has been systematically questioned by Landau and Hunt (1990) and the ensuing 'Rhizotomy Correspondence' (1991) together with reviews by others not actively involved in

a rhizotomy programme(Neville,1988 and Bleck,1993).

i. Physiological basis for (SDR).

Unlike IT baclofen, there have been no reported deaths using SDR. In common with the baclofen rational, the use of SDR is aimed at reducing posterior root-mediated spasticity. However, it has been principally targeted at the child with cerebral diplegia who has already attained walking mobility, is free from cognitive impairments or seizures, has well-motivated parents, access to good physical therapy facilities and shows no evidence of extrapyramidal manifestations (Peacock and Staudt, 1990; Park and Owen,1992).

The 'rationale' for SDR is to reduce 'spasticity' by depriving the central nervous system of the afferent input supporting it is based on the original observations of Sherrington (1988) that dorsal rhizotomy relieved 'spasticity' in cats and Foerster's clinical application of the technique in man. Foerster (1913) used electrical stimulation to identify and cut the posterior roots from L2 to S2 spinal afferent rootlets, sparing the L4-5 roots to preserve quadriceps muscle strength for standing. Foerster (1913) drew attention to the need for a prolonged postoperative exercise regimen and the subsequent need for orthopaedic surgery in many cases. Despite treating some 159 cases, 88 of whom were described as suffering from "congenital spastic paraplegia" and 8 cases with "infantile spastic paraplegia", the technique saw little use till it was revived by Gros *et al* (1967), who modified the technique by restricting division to four fifths of the rootlets from L1-S1 spinal segments. Gros and his team attempted to identify rootlets making connections with useful muscles such as the abdominal muscles, gluteus maximus, quadriceps femoris and gastrocnemius muscles, which he spared, while dividing rootlets supplying adductors and hip flexors. The 18 year long-term follow-up of 62 case by Gros (1979), indicated a 25% failure rate which was attributed to errors of selection. Complications included a third (22/62) of cases with hypoaesthesia in the limbs, but in the 25 cases with cerebral palsy, additional benefits reported included improvements in language and upper limb function.

Another modification of the rhizotomy technique was developed by Fasano and colleagues (1978, 1979, 1988) for use in cerebral palsy patients which involved selection of the dorsal rootlets by intra-operative electrical stimulation: this technique being adapted by Peacock and Arens (1982) who also lowered the level of the laminectomy from L2-L5 to enable exposure of the cauda equina, identification of L2-S2 spinal rootlets which thus

avoided inadvertent division of S3 rootlets perceived as important for bladder sensation and control.

ii. Selection of posterior rootlets by intra-operative electrical stimulation or at random?

According to Fasano's new approach (1979, 1988) for the selection of suitable posterior rootlets for division, a train of electrical stimuli applied to the dorsal rootlets produced an unsustained contraction if the stimulated rootlets did not directly support the spasticity, whereas a sustained or tonic discharge and contraction occurred in rootlets supplying spastic muscles. Thus, the rootlets associated with brief discharges were spared and those producing sustained discharges were cut.

There have been a number of studies examining the neurophysiological basis of the intra-operative rootlet selection by electrical stimulation.

Laitinen *et al* (1983) performed SDR in 9 adult patients:6 cases of multiple sclerosis, and three with cerebral haemorrhage, spinal injury and unspecified myelopathy respectively. One case had rhizotomy at the C6-C8 cervical level, 5/9 cases were rhizotomised between T12-S1 roots and 3/9 cases underwent L1-S1 rhizotomies. The authors reported that 60%-80% of the stimulated fascicles responded with sustained tonic muscular contractions and accompanying surface EMG discharges when exposed to trains of stimuli at a frequency of 60Hz, with a stimulus duration and amplitude of 0.2ms and 1-6V respectively, applied for between 1-3s. These fascicles were then divided, abolishing spasticity in 4/9, markedly reducing it in 3/9, and partially reducing it in 2/9 cases respectively.

Pollack (1994) has reviewed the intra-operative stimulation technique, classifying the responses from 1-4 as follows:

1. single muscular contraction
2. continuous muscular contraction
3. spread of contraction and EMG activation to ipsilateral myotomes removed from the root stimulated.
4. Widespread ipsi- and contra-lateral leg muscle EMG discharges and contractions.

Several short-comings of the method are cited: the first being that type 1 responses are seldom reported and the second problem is the reported occurrence of a type 2 (tonic response) in individuals without clinical evidence of spasticity (Cohen and Webster, 1991; Phillips and Park, 1989). Even when a statistical framework for evaluating the stimulation

responses is applied, Pollack (1994) concluded that the benefits of intra-operative stimulation were minimal, questioning the additional cost of such a procedure over a random selection of rootlets, as well as highlighting the fact that the ventral (motor) roots have a lower stimulation threshold than the dorsal roots, potentially allowing more rootlets to be mistakenly divided on the basis of direct muscle stimulation rather than the intended reflex muscular stimulation.

Underlining the difficulties imposed by anaesthesia, Logigiano and colleagues (1994) found that the H-reflex was unobtainable in 5/10 cerebral palsy cases some 120-180 minutes after onset of anaesthesia. One of their major concerns was the relative ease with which the ventral roots could be inadvertently stimulated to produce surface EMG discharge patterns at a rate of 50Hz that looked indistinguishable from an H-reflex.

The H-reflex suppression by intra-operative anaesthesia and ease of direct muscle stimulation raises serious questions about validity of the intra-operative stimulation and the 'selectivity' of procedure in identifying dorsal rootlets for division.

iii. Follow-up studies of selective dorsal rhizotomy in children

This treatment has become routine in North America (Oppenheim, 1992) and children are being offered treatment at increasingly young ages as recommended by Park and Owen (1992). however, as indicated above, the intra-operative selection procedure has been shown to over-diagnose the presence of afferent rootlets 'mediating spasticity' because in many cases, the muscles are being stimulated directly along the motor nerve and not reflexly as had been supposed. Random division in the majority of centres performing the operation seems to be the norm (see Steinbok, 1997), although a group in Seattle, Washington, attempted to use the stimulation technique in their study (McLaughlin *et al*, 1996).

Neurophysiological assessments before and after SDR are scarce. Cahan and colleagues(1987) reported the results of pre- and postoperative cutaneous somatosensory evoked potentials (SSEP) H-reflex and Hmax/Max ratio studies in 20 children undergoing SDR. They found that:

1. 9/20 children had abnormal posterior tibial SSEP pre-operatively, in 8/9 of these, the median nerve SSEP was normal, indicating that preoperative evidence of spinal cord dysfunction exists in some children
2. surgery led to loss of the posterior tibial SSEP in 3/11 cases

3. in 6/9 cases with preoperative abnormalities of the posterior tibial SSEP, the SSEP waveform improved postoperatively
4. the Hmax/Mmax ratio decreases post operatively in keeping with tone reduction in 7/8 cases and an increase in the ratio in 1/8 cases.

Lazarreff and colleagues (1990) reported on 30 children aged 4-12 years with 'spasticity" who underwent rhizotomy and whose spasticity severity was graded on a 5 point scale (table 1.3.5.3.). The laminectomy performed was from L5-S1, the L4, 5 and S1 roots identified and stimulated and the muscle responses observed visually and registered by EMG: division of rootlets was then administered to those exhibiting a sustained contraction or prolonged EMG discharge as described above. Overall 40% of stimulated L4 and 50% of L5-S1 rootlets were divided. The patients were assessed 1 week before, within one week post operatively and 6 months later.

Table 1.5.3. Spasticity grading after selective dorsal rhizotomy

Spasticity	Score	Description
Absent	0	none
Mild	+	No abnormal posture Normal passive movement No disability
Moderate	++	Abnormal posture: completely reduced by passive movement Full joint range of movement Higher muscular tone than in higher grade Moderate disability
Marked	+++	Abnormal posture: incompletely reduced by passive movement Limited range of joint movement High resistance to muscle stretching Marked disability
Severe	++++	Abnormal posture slightly reduced by passive movement Severe disability.

after Lazarreff et al, 1990

The results indicated a similarity in the responses to testing from both sides of the body with apparent improvement in the above scoring system in the upper-limb muscles, particularly wrist flexors, and for the lower limb, the 'spasticity score' of the lower limb muscles showing the greatest reduction moving from 'moderate' or 'marked' categories to the absent or mild 'groups'.

However it is not clear if this picture was maintained at 6 months or simply refers to the

first post-operative week. Furthermore, the postoperative ambulatory status reflected the pre-operative ambulatory status, only three cases being independent and 9 nine orthotic-dependent ambulators pre-operatively. The majority of children (21/30) being non-walkers. Lazarreff *et al* report 8 cases who initiated supported walking and a further nine who progressed to floor creeping and the remaining five who attained improved length of sitting post-operatively. In addition improved self-feeding, using the telephone, hand grasp and handwriting are reported, whereas no cases were hypotonic and there were no reports of increased foot-valgus deformities reported by others (see below: complications of rhizotomy).

Cahan and colleagues (1990) reported the results of 14 cases who underwent SDR and had pre- and post-operative gait analysis: the surgery removed the spasticity and abolished clonus of calf muscles at foot-contact in the 7 cases who were independent ambulators, as well as improving the dynamic spatio-temporal parameters of gait such as increasing the walking velocity by 42% and stride length by 35%, while cadence remained unchanged: however the pattern of muscle activation during walking remained unchanged, although 1/6 cases who had required assistive devices progressed from supported to independent walking . The authors also conclude:

> "The frequency of postoperative valgus and excessive dorsiflexion suggests that an orthosis should frequently be prescribed postoperatively to control hind-foot inversion/eversion and to limit dorsiflexion."
>
> *Cahan et al, 1990.*

Vaughan, Berman and Peacock (1991) reported gait analysis studies three years post rhizotomy in 11/14 cases who had been pre-operative ambulators, showing an improved knee joint range of motion although thigh motion was initially above the normal range, returning to the normal range at the end of the three years. Stride length and walking velocity were said to have improved, though cadence re,remained unchanged.

Peacock and Staudt (1991) also report the results in 27/42 cases who had previously undergone SDR of 20% to 50% of stimulated rootlets. 8/27 cases were classed as spastic diplegia, who were independent ambulators without orthotics or assistive devices; 14 were 'spastic diplegics' or quadriplegic patients who walked with assistive devices or support; two cases had 'spastic quadriplegia' and could creep and three were severely affected, of which only one was included in the study as it was impossible to obtain accurate measurements in

the other two. Overall there were 25 cases who underwent formal evaluation (mean age 5.9, range 3.25-10.25 years). Assessments were performed in the week prior to surgery and 5-14 months post-operatively (mean 8.9 months), and the results indicated statistically significant improvements in spasticity scores (based on a combination of Ashworth (1964) and Bohannon-Smith (1987) scores, with abolition of clonus and depression of deep tendon reflexes. In lower limb, but not upper limb, joint ranges were likewise improved. However muscle strength was reduced post-operatively in thigh abductor and foot dorsiflexor muscles.

Function scores in this cohort showed improvements in several of the 16/25 children whose function was assessed: nine scored higher in sitting, seven in standing, two in crawling and seven in walking. One child whose standing and walking grade declined and another with a decline in crawling grade recovered their preoperative ambulatory status after longer follow-up. In the remaining nine cases, improvements were perceived in static postures and transitional movement skills: 5 cases requiring less support in half-kneeling, six with improved transition between kneeling and half-kneeling and four with improvements in rising to stand from sitting. Two children underwent orthopaedic surgery for pre-existing contractures.

A prospective follow-up in an observational study of 34 children with "spastic CP" (24 diplegic, 10 quadriplegic), while showing a reduction in spasticity as judged by the Ashworth Scale and deep tendon reflexes, showed "considerable variability" in outcome. (McLaughlin *et al,* 1994).

In a preliminary report of a randomised control study of 24 children, 12 of whom were randomised to SDR, there were no statistical differences in the gross motor function scores at 12 months after SDR (Wright *et al*, 1994): while improvements in stride length, stride time and velocity were noted, the significance of these findings was weak (Shiel *et al*, 1994). Another group (Thomas *et al*, 1994) reported no differences in gait parameters 1-2 years postoperatively.

There have been two recent randomised controlled studies with blinded evaluation to compare the effects of SDR+physiotherapy with physiotherapy alone.

i. Steinbok and colleagues (1997) studied 28 children with diplegia at baseline and 9 months after SDR+ Physiotherapy or Physiotherapy alone and found significant

improvements in the clinical mobility scores, Gross Motor Function Measure(GMFM), Ashworth measure of "spasticity" and joint ranges in children who had undergone SDR. However, there was no improvement in the physiological cost index between groups.

ii A similar, but slightly larger study based in Seattle, Washington, USA (McLaughlin and colleagues, 1996 awaiting formal publication) failed to show any differences in GMFM between SDR+PT(19 cases) and PT alone (19 cases) when evaluated at 12 months. This study also employed the intra-operative electrical stimulation of the dorsal afferent rootlets for rootlet selection advocated by Peacock and Staudt (1990) and Oppenheim et al (1992), however the likelihood is that this technique is flawed, resulting in a direct M-response from co-stimulation of ventral efferents rather than producing a train of H-reflexes (Soriano *et al*, 1995; Logigian *et al*, 1994).

iv. Complications of SDR.

Weakness is a common feature in cerebral palsy (Brown et al 1987, 1991), though may be overlooked. As indicated above, Peacock *et al* (1991), weakness is a clearly measurable deficit postoperatively and an absolute requirement for an active post-operative exercise regimen has been highlighted Giuliani (1991).

A number of studies reveal the return of spasticity in as little as a year from SDR and the need for repeated orthopaedic interventions (Bretas *et al,* 1991; Sienko *et al*, 1994). Specific complications include: Excessive dorsiflexion and valgus requiring orthotic correction (Cahan *et al*, 1990); spondylolisthesis and scoliosis from laminectomy (Peter *et al*, 1990); rapid hip subluxation (Green *et al,* 1991) and acquired vertical talus (Mooney *et al*, 1994).

v. The place of SDR for children with cerebral palsy.

The basic assumptions underlying the pathogenesis of the movement disorder in 'spastic diplegia', the case selection for SDR, and selection of the rootlets for section during the operation have been systematically questioned by Landau and Hunt, 1990, ensuing 'Rhizotomy Correspondence' (1991) and more recently by the reports from Pollack (1994) and Logigiano *et al* (1994). In only one of the reported studies (Cahan *et al*, 1990) was there an attempt to measure velocity-dependent stretch hypertonus, in this, and all the other studies above, including the recent randomised studies, a *syndromic* approach to classifying the cases was adopted.

One overt aim of rhizotomy is to treat spasticity while at the same time preventing the need for multiple orthopaedic procedures over many years. The current scientific evidence for the basis of rootlet selection and benefit of SDR is conflicting and unresolved, despite widespread espousal of the technique in North America. Nor is it entirely clear that treated children require less in the way of traditional therapies such as orthotics and local tendon and bone surgery.

The history of dorsal rhizotomy-total, selective, and random- illustrates the current problems with basic physiological concepts, case definition and outcome measures to evaluate the effectiveness of therapy over and above the natural history of the motor disorder as described in section 1.4, above.

1.5.4. Botulinum toxin A.

As has been seen, treatments with intrathecal baclofen or selective dorsal rhizotomy present considerable difficulties for case-definition, selection and follow-up which, combined with the medical investment in manpower and equipment required to deliver such services significantly limit their clinical usefulness.

The use of botulinum toxin A, which began with the treatment of blepharospasm laryngospasm and torticollis in patients suffering from dystonia, appears ideal for the management of dynamic deformities and postures. Problems with design methodology and objective measurements in the seven published studies on children receiving botulinum toxin A have been reviewed by Forssberg and Tedroff (1997), who highlight some of the pitfalls in research methods in the whole field of cerebral palsy rehabilitation.

Botulinum toxin A inhibits the release of acetylcholine from the presynaptic nerve ending of the neuromuscular junction. This effect reaches a maximum over a few weeks but lasts several months. It has the potential benefit of being repeatable if and when signs recur.

An additional advantage is that since the muscle is the final common pathway of all motor activity, botulinum toxin A abolishes or reduces voluntary and involuntary movements, dystonia, tonic labyrinthine reflexes, spasticity and reflex excitability in the injected muscles. It is therefore suitable for the treatment of dynamic hypertonus of any origin. However, by definition , treated muscles will become weak, if not paralysed.

Whether or not Botulinum toxin A prevents contractures as was demonstrated in the 'spastic mouse' (Cosgrove and Graham, 1994) or in early reports in children with dynamic

equinus, remains to be fully established, though early studies of its use in the lower limb for dynamic equinus (Cosgrove, Corry and Graham, 1994; Sutherland *et al*, 1994) and for use in the upper limb to improve hand cosmesis (Corry *et al*, 1997) appear promising.

This chronic "poisoning" of the muscles is a relatively new form of treatment but botulinum toxin A does provide the ability to tailor treatments individually at relatively low cost in a way which proves impossible with conventional orthopaedic surgery or rhizotomy.

The risk of systemic poisoning with respiratory failure is small but nonetheless exists, necessitating caution particularly in cases in whom respiratory function is already compromised by poor coordination and spinal deformities.

1.5.5. Dilemmas in the Clinical Management.

The needs of a child with CP may range from being indistinguishable from the normal child to total dependency with a developmental age fixed at less than 3 months. Treatment depends on:

1. The expectations of the parents and the child
2. Knowledge of the natural history of cerebral palsy phenotypes
3. The mechanisms of action of different management strategies
4. Adequate outcome measures
5. Availability of resources and expertise to deliver services.

The first objective is to retain the trust of the parents and to be open and honest about problems. In infancy, it is essential to avoid the possibility of the child being rejected. Rejection may lead to the child being abandoned to fostering, and denial, to unrealistic expectations, anger and frustration.

It is all too often the case that the child is too young to be consulted at a time when radical treatments such as orthopaedic surgery are carried out. As the children become adults, their needs are often neglected. It is vital to bear in mind that the "natural history" of CP may also be acting in the child's favour in terms of slow, but continued development.

If a child can sit independently at the age of 2 years, they are likely to progress to standing and walking, though they may need orthotics (splints and crutches) and exercise tolerance may be extremely limited. Also, the likelihood of attaining ambulation is 100% for all hemiplegic children, diminishing by about 10% for each additional limb involved to 70% in quadriplegia in the spastic syndromes which comprise the majority of cases of CP. The onus

on carers, is to ensure as far as possible that the child can benefit from this slow development.

At present , there is insufficient understanding of the pathophysiology of CP for prescriptive treatment. Current methods of assessment poorly predict the likely effects of any given intervention.Treatments and their evaluation remain crude, often relying on one-off procedures to alter a dynamic and continuous process. It should be remembered that CP is a disorder of motor control, ie a movement disorder due to abnormal muscular activation patterns producing abnormal movements (Leonard *et al*, 1991) and postures (Nashner 1985).however, goal-directed physiotherapy (Bower and McLellan, 1992) as well as intensive exercise training are proving to be strong contenders to the 'ablative therapies'.

The issue of mobility is particularly complex and important for children at several levels and helps to focus beyond considerations of mere impairment of a given limb or limbs to include vision, a sense of direction, a sense of danger to self and others, of time (arriving early or more usually late) of energy demands and economy and finally the complex fusion of choice and desire. Mobility therefore equates with independence and control over one's surroundings. Often, a distinction must be made between home and community mobility. At home the child may continue to crawl or walk with a rolator or other aids but community mobility may be impossible without the aid of a wheelchair which may need to be powered.

The child who learns to walk or to steer a wheel-chair, is using visual skills as well as demonstrating visual awareness. It is a common experience to see overprotected children who by dint of being passively transported in their chairs behave as passengers in a vehicle: they look about in an undirected fashion and so fail to develop the skills of looking for obstacles which becomes second nature when one is self-mobile.

1.6. Summary.

i. This selective review of the cerebral palsies has attempted to convey the inherent complexities of the neurological presentations in cerebral palsy with a view to highlighting future directions for prevention and management.

ii Infarctive versus non-infarctive white matter pathologies have been reviewed, generally reflecting the birthweight or gestational age of the infant and different pathogenetic mechanisms of injury, indicating likely avenues for prevention of brain injury.

iii Problems relating to the relationship between structure and function in the nervous

system and to dysfunction following injury have been touched upon with emphasis on neurological changes within the central nervous system distal to the site of the injury, indicating the likelihood of spinal cord dysfunction in children with cerebral palsy.

iv Of particular importance, has been the emphasis on the early prognostic indicators for ambulation and the lack of progress in modifying the inherent natural history of cerebral palsy over the last 40 years.

v The neurological sub-types and the confusion over the use of the term 'spasticity' to mean both a syndrome and an operationally-defined pathophysiological variable has been emphasised repeatedly with particular reference to the natural history of cerebral palsy and the use of specific 'antispastic' therapies.

vi Finally, three invasive and controversial treatments have been reviewed because of their impact on clinical practice today in comparison with more widespread use of physical therapy, orthotics, soft tissue and bony surgery and oral tone-relieving agents.

In the following sections, the physiology of muscle tone will be reviewed (section 2) followed by studies on the mechanisms of equinus (section 3); of the non-reflex / non-electrical hypertonus of plastic muscles (section 4); reflex excitability in thigh (section 5) and calf (section 6) muscles; the relationship between soleus reflex excitability, reflex muscle twitch and the joint angle in adults and children (section 7) and in children with hemiparesis (section 8) is extensively studied with insights on the effects of common treatments such as casting and tendon surgery in relation to the phenomenon of ankle clonus.

Section 9 deals with the physical laws controlling alternating movements and some of the factors contributing to the maturation of motor dexterity.

Section 10 dwells on the importance of recognising the difference between posture originating from tonic labyrinthine and tonic neck reflexes and spasticity *per se*, together with a demonstration of the effects of sleep in abolishing the tonic labyrinthine reflex and spontaneous extensor toes. Associated movements are briefly reviewed.

Section 2. The Physiology of Muscle Tone.

2.1 Definition (s) of Muscle Tone

"Muscle tone" is frequently identified as a clinical problem but hardly ever defined. Some authors (Fenn and Garvey, 1934) have adopted the view that the term "muscle tone" is essentially meaningless without qualification. Brodal (1981, p 166) summarises the clinical approach to evaluating "muscle tone":

"Muscle tone. If a normal muscle is palpated when it is resting, it will be felt that it is not completely flaccid but possesses a certain degree of tension . This is also the impression gained when passive movements are made. This normally existing condition of muscular tension is generally called muscle tone, more properly resting tone. In various pathological conditions it is observed that this normal tone is changed, sometimes increased (hypertonus), in other cases reduced (hypotonus). Clinically, muscle tone is commonly examined by palpation and passive movements. However, probably two different components of muscle tone are evidenced by the two methods of examination (see below). Thus it is not uncommon to observe, for instance in capsular hemiplegias, that muscle tone is reduced when judged by palpation (the consistency of the muscles is reduced), while in passive movements it appears to be augmented, since the resistance offered to the movements is greater than normal."

Brodal (1981), p 166

One of the major difficulties in clinical practice has been in establishing clear and unambiguous operational definitions of "muscle tone". The usefulness of operational definitions is clearly evident in other medical disciplines where they have contributed to medical advances, an obvious example being the scalar variable "blood pressure". The tools required for its measurement, such as an inflatable cuff connected to a manometer or strain gauge and a stethoscope; the method(s) for using the tools can be specified: ie occlusion of the artery in a specified limb by inflating the cuff which has been wrapped round the limb until the heart sounds in the artery distal to the occlusion are effaced or, more commonly return on gradually allowing the cuff to deflate. Such a simple method has opened up the fields of cardiology, nephrology, endocrinology, neurology and psychology, since all of these organ systems may influence the scalar quantity known as "blood pressure". Once so defined, refinements in technology inevitably follow along with clinical methods of standardising measurements eg by specifying how the recordings should be taken, for example, from the non-dominant arm, the leg, via an arterial catheter, or while lying, sitting or standing, during or after exercise or a prescribed period of rest: in other words, the internal and external

influences which may be brought to bear on the phenomenon in health and disease may be investigated. For an extensive instrumented study on "muscle tone" see Lakie, 1981, and Walsh, 1992.

Unlike simple scalar quantities such as mass, molar quantity, temperature or blood pressure, the anatomical and physiological variables contributing to the mechanics of "muscle tone", involving combinations of mass, length and time (ie vectors), are more difficult to study but may be divided into those factors which are mediated by active muscle contraction (electrically active) and those which stem from the passive (electrically silent) properties of the muscles and tissues:

> "There has been much debate on the problem of muscle tone, some confusion has arisen because various definitions have been employed. The resistance offered by a muscle to being stretched may in principle be due to two factors: the inherent viscoelastic properties of the muscle and the tension set up by contraction. Both factors are clearly important, but their respective components obviously differ in various situations."
>
> *A. Brodal 1981, p166.*

Such an extension of the clinical method is shown in table 2.1, together with some of the defining characteristics of the variable which indicate the appropriate conditions under which the variable may be studied. Accordingly, the resistance to passive stretch, which is felt as the limb is put through a full range of movement about a joint with the patient relaxed, is an amalgam of biomechanical and neurophysiological factors affecting the the state of the muscle (contractile elements) and soft tissues (tendon, fascia, joint capsules etc.,).

2.2 Biomechanical components of muscle stiffness.

As indicated in table 2.1, muscles possess elastic, viscous and inertial properties which are displacement, velocity and acceleration-dependent phenomena respectively. For the purpose of discussion, it is customary to refer to these various biomechanical components of muscle and tendon as collections of in-series and in-parallel springs and "dashpots", figure 2.2, (see Rack *et al*, 1983).

Table 2.1. Components of Muscle Tone: (resistance to muscle stretch).

I. Biomechanical (non-electrical): contributions from passive rheological properties

Physical Variable	Site Generated	Operational Definition
1. elasticity	intramuscular	length-dependent
2. viscosity	intramuscular	velocity-dependent
3. viscoelasticity	intramuscular	time-dependent
4. inertia	intramuscular	acceleration-dependent
5. friction	intramuscular/joint	independent of length or velocity
6. plastic*	intramuscular	time-dependent
7. contracture	intramuscular	short muscle / short tendon

II.Neurophysiological (electrical): contributions from active muscle contractions

Clinical Variable	Site Generated	Operational Definition
1. Hypotonia	Intramuscular** neuromuscular jctn neural CNS	a general absence or reduction of muscle activity which may be congenital acquired, continuous, intermittent or sudden, depending on mechanism.
2. Myotonia	intramuscular	slowness to relax:decremental EMG
3. reflex excitability and #spasticity	reflex arc/supraspinal	velocity-dependent: Muscle silent at rest, abnormally low reflex velocity threshold and increased reflex gain to stretch.
4. dystonia	CNS	Muscle continuously active producing fluctuations in muscle tone with changes in labyrinthine input, body contacts and non-specific afferent inputs: abolished by sleep.
5. Parknisonian rigidity	CNS	A state of increased resting muscle activity: abolished by sleep but not deafferentation associated with typical tremor, akinesia and hypokinesia, and exaggerated long-latency stretch reflexes.
6. posture	CNS	Tonic neck reflexes Tonic labyrinthine reflexes: eg hemiposture Associated postures: eg Føg and mirroring voluntary and involuntary
7. movement	CNS	voluntary and involuntary.

*This includes thixotropy, creep and stress relaxation.

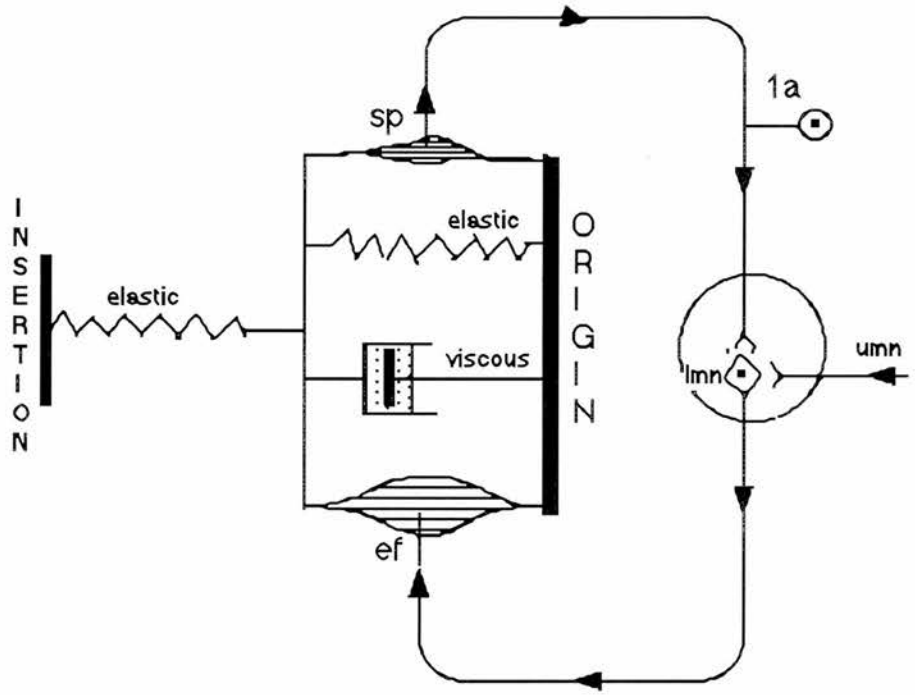
** The mechanism of hypotonia is largely unknown: see Struppler 1987 (below) for intramuscular mechanism (s) post thalamotomy.

As defined by Lance, 1980; see section 1.3.3 for "syndromic" ie non-operational definition

Figure 2.2. illustrates the biomechanical and neurological factors resisting stretch.

Figure 2.2. Schematic diagram of the in-parallel arrangements of the contractile elements of the extrafusal muscle fibres (ef) and the intrafusal muscle spindle sense-organs (sp), together with the sensory 1a afferent loop from spindle to spinal cord, the lower motorneurone (lmn) innervating the extrafusal fibres (ef) and the supraspinal upper motor neurone (umn). The elastic elements, drawn as springs are both in series and in parallel with the muscle; the viscous element, drawn as a 'dashpot' lies in parallel with the muscle. Note: the spindle (sp) and extrafusal fibres (ef) also exhibit time-dependent 'plastic 'properties such as 'thixotropy', 'stress relaxation' and 'creep' (qv). (The gamma motor neurones innervating the muscle spindle and the supraspinal 1a projections are not shown. For role of golgi tendon organ, 1b afferents and interneurones see fig 2.3)

drawn after Rack et al 1983



2.2.1 Elastic resistance to stretch.

'Elasticity' is defined (Chambers Materials Science and Technology Dictionary, 1993, p105) as:

"the tendency of matter, whether gaseous, liquid or solid, to return to its original size or shape, after having been stretched, compressed, or otherwise deformed."

Elastic properties can be measured using methods which examine the relationship between length and tension according to Hooke's principle (Wright and Johns, 1960; see Walsh 1992, pp 52,83, 172) and is illustrated in figure 2.2.(after Wright and Johns, 1960).

Muscle is a complex tissue with essentially non-linear characteristics, nevertheless, the concept of elastic properties is useful, especially in pathological states dominated by muscle contracture when the contractile components of muscle may be considerably reduced (see below) and the muscle-tendon complex behaves in a more spring-like manner. In physical terms, the elasticity of any given material is given by Young's Modulus which is determined by the 'stress' to 'strain' ratio:

$$\textbf{Young's Modulus of Elasticity} = \frac{\textbf{Stress}}{\textbf{Strain}} \qquad 1.$$

where **Stress** = $\frac{\text{force applied}}{\text{area of material}}$

and **Strain** = $\frac{\text{deformed length}}{\text{original length}}$

Accordingly, Young's Modulus of Elasticity = $\frac{\text{force applied X original length}}{\text{area of material X deformed length}}$ 2.

The spring-like properties of muscles probably do serve to store up energy and absorb shocks but if muscles were anything like pure springs, the body segments would oscillate intolerably and the resulting motions would be uncontrollable, the inherent braking system being the viscous element, acting to dampen, restrain and smooth intended and unintended motion.

In a study of reaction times, Winter and Brookes (1990) looked at the 'premotor time' and 'electromechanical delay'. The premotor time was defined as the interval between application of a stimulus and the change in electrical activity (EMG) of recruited skeletal muscle . 'Electromechanical delay' (EMD), being the interval between EMG onset and movement. Previous studies had indicated most of this EMD interval being taken up by the time to stretch the series elastic component in muscle (Alexander and Bennet, 1977). Winter

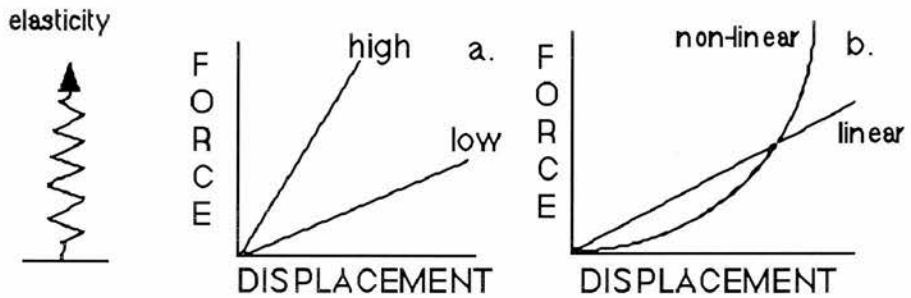
and Brookes (1990) were able to arrive at the following computations of 10 voluntary soleus contractions in response to an acoustic signal in 13 healthy adult subjects,(table 2.2.1):

Table 2.2.1 Electromechanical response times (ms)of the soleus muscle (n=13).

Variable	Mean	SD
Total reaction time	171.6	26.0
Pre-motor time	130.5	24.8
electromechanical delay	40.8	4.8
force time	9.5	3.3
elastic charge time	31.5	4.9

after Winter and Brookes 1990

Note that 'force time' denotes the interval between EMG and onset of the muscle tension, while the 'elastic charge time' is the interval between onset of muscle tension and movement.



Elastic stiffness:
a. linear relationship between force and displacement, elastic stiffness given by the slope .
b. linear and non-llinear elastic stiffness.

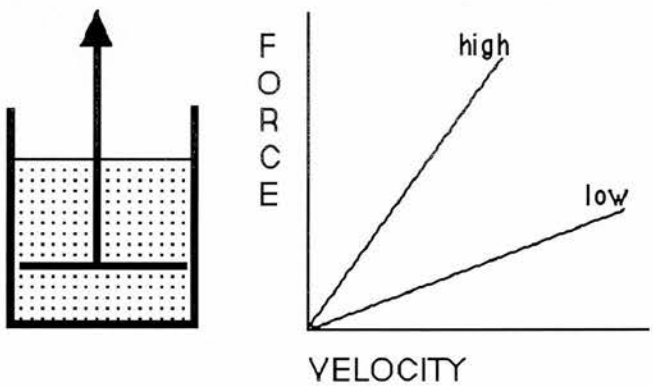
Figure 2.2.1 Linear and non-linear models of elastic stiffness given by the slopes of the force:displacement plots. *after Wright and Johns, 1960*

2.2.2 Viscous resistance to stretch.

Strictly speaking, the term "viscosity" applies to liquids, but as we shall see, muscles do behave in part as special liquids. Viscosity is defined as:

"The resistance of a fluid to shear forces, and hence to flow. In a Newtonian fluid, such shear resistance is proportional to the relative velocity between two surfaces on either side of a layer of fluid, the area in shear, the viscosity of the fluid and the reciprocal of the thickness of the layer of fluid."

Chambers Materials Science and Technology Dictionary, 1993, p330



Viscous stiffness: measured by plotting force against velocity, the higher the viscosity, the steeper the slope of the relationship.

Figure 2.2.2 Physical behaviour of an ideal viscous material.
after Wright and Johns, 1960.

The viscosity, or coefficient of viscosity is defined by:

$$\textbf{Coefficient of Viscosity} = \frac{\textbf{Shear Stress}}{\textbf{(dy/dt)}} \quad 3.$$

Where $$\textbf{Shear Stress} = \frac{\textbf{Shear Force}}{\textbf{Area of action}} = \frac{\textbf{F}}{\textbf{A}} \quad 4.$$

and **(dy/dt)** =**Shear Strain Rate**, or **velocity gradient**

giving $$\textbf{Coefficient of Viscosity} = \frac{\textbf{Shear Force}}{\textbf{Area X Velocity gradient}} \quad 5.$$

Hence, broadly speaking, viscosity is inversely proportional to the velocity and the force required to overcome viscous resistance is directly proportional to the velocity employed. The analogy often used is the experience of dragging a spoon through a pot of honey: the ease of progress diminishes with the speed of the spoon; similarly, when attempting to walk through water, it is easier to walk slowly than at speed. To measure the 'viscous" component of resistance to stretch, the muscles must be stretched at different velocities, although, in fact, muscles would be classed as a rather special case of a 'viscoelastic' material.

2.2.3 Viscoelastic resistance to stretch.

According to the Chambers Materials Science and Technology Dictionary (1993, p332):

"The response of viscoelastic materials to an applied load is a combination of viscous and elastomeric components. The viscous component in polymers is caused by chain slippage in the amorphous zones and is time-dependent and irreversible. The elastomeric component arises in the same zones but is reversible, being caused by entanglements trapping chain segments which strive to return to their equilibrium position when unstressed."

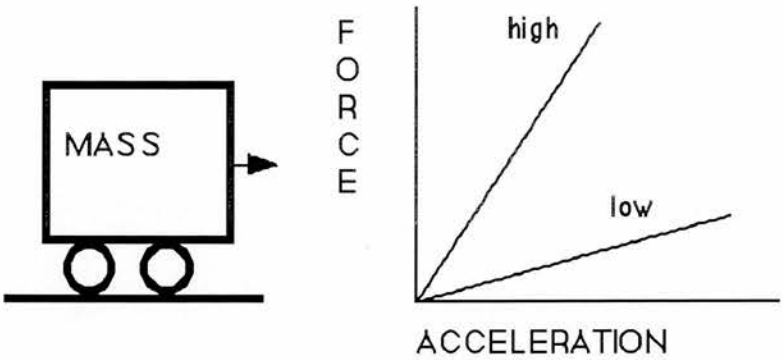
Four types of viscoelastic behaviours are noted:

a. glassy-brittle
b. leathery
c. elastomeric
d. viscous

A dramatic example is given of a material capable of exhibiting all four behaviours under different circumstances:

" All four viscoelastic regimes are shown rather dramatically by 'potty putty'. a low molecular mass silicone rubber cross-linked with boron oxide. By the time-temperature equivalence principle, increase of temperature is equivalent to increase of time scale; so when hit with a hammer, the material shatters like glass. When rolled into a ball and dropped, it bounces like rubber but yet it can be kneaded like putty with the fingers. At long time scales it flows like a viscous liquid and will cover a wide area without any applied force. Viscoelastic behaviour is characteristic of polymeric materials and is shown by asphalts, bitumens, paints, foods and many biomaterials such as wood and bone."

Chambers Materials Science and Technology Dictionary (1993, p332).



Inertial stiffness:
The larger the mass, the greater the inertia, the greater inertial stiffness. Inertial stiffness given by the slopes of the force:acceleration plots

Figure 2.2.4. Inertial Stiffness. Ideal representation of a mass on a frictionless surface.
after Wright and Johns, 1960.

2.2.4. Inertial resistance to stretch.

The inertial resistance to movement (fig. 2.2.4, above) is of course made up of the muscular and skeletal contributions to limb mass. The arrangements and lengths of the long bones of the upper and lower limbs dictate, to a considerable extent, the types of motions possible at each joint. During passive and active motions at any given joint, inertial resistance must be overcome by appropriate torques, ie forces acting on levers about a joint. The moment of inertia is given by the sum of the the unit mass of the segment multiplied by the square of the lever arm of each unit mass, given by the relation:

$$\textbf{Moment of Inertia, } I = \Sigma mr^2 \qquad 6.$$

The torque, **T**, required to overcome the inertial resistance is given by the moment of inertia, **I**, multiplied by the angular acceleration, **a**:

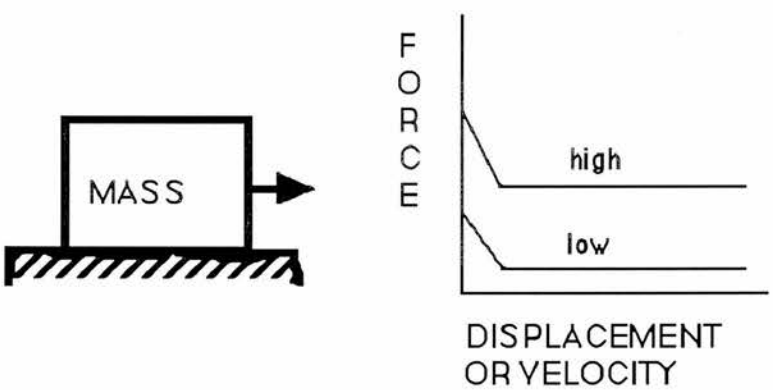
$$\textbf{Torque} = Ia \qquad 7.$$

The relationship of inertial resistance with limb length is interesting and has been

studied by Walsh and Wright (1987, fig 3, p 165). The inertia of the forearm was shown to have a linear relationship with the *fifth power* of the limb length, the lowest inertias being recorded, as might be expected, in the two paediatric subjects. The significance for motor control as the child grows into an adult is significant, since a doubling in limb length results in a thirty-two fold increase in limb inertia which must be matched by an equivalent increase in muscular force. The inertial contributions to resistance to motion may be studied under isotonic conditions at different angular accelerations. The contribution of inertial elements to the physical problems of voluntary movements is explored in section 9, below.

2.2.5 Frictional resistance to stretch.

Wright and Johns (1960) were unable to find evidence of significant frictional resistance to stretch at the interphalangeal joint of normal subjects in which the frictional resistance represented less than 1% of the total torque. By contrast, elastic stiffness accounted for 90% and viscous stiffness, 9% of the total torque applied to move the quiescent finger. Frictional resistance is depicted in figure 2.2.5.



Frictional stiffness:
The force required to overcome frictional stiffness is initially high then becomes constant and is independent of displacement or velocity.

Figure 2.2.5 Frictional resistance to stretch. *after Wright and Johns, 1960.*

Each of these viscoelastic and inertial resistances may be separately analysed under suitable conditions by non-invasive means. However, it is important to recognise that the above definitions relate to idealised properties of muscle, tendon and bone, since in reality, these tissues exhibit considerable 'plasticity' (see 2.2.6).

2.2.6 Plastic muscle properties.

As indicated in the foregoing sections, tissues such as muscle, tendon and bone exhibit plastic properties which can be studied under different conditions. From a medical viewpoint, 'thixotropy', 'creep' and 'stress-relaxation' are particularly interesting.

2.2.6 1. Thixotropy.

Muscle tissue exhibits time-dependent "plastic" properties such as "thixotropy" (Lakie, Walsh and Wright, 1984a and 1984b). Walsh (1992) discusses the effect of "touching" or "stirring" muscles:

"Stiffness that depends on the past history of movement characterises thixotropy (from the Greek words 'thixis' or 'touch' and 'tropos' meaning 'turning')."

"The term was introduced by Péterfi (1927), who noticed changes on stirring, with a needle under a microscope, the cytoplasm of sea-urchin eggs. The gel turned to a sol but reverted to being a gel after a period when it was not disturbed. This is a common property of large molecules in solution. Agitation disrupts bonds which form between the molecules and which take time to reform once the motion ceases. This behaviour is found in materials such as clays, paints and sauces. For tomato ketchup there is a ditty' 'it's shake and shake the ketchup bottle, none will come and then a lot'll'. Sometimes thixotropic compounds are used in pharmaceutical preparations. Bentonite is added to calamine lotion to improve its physical properties, so that it does not run off the skin. The chemist's instructions to 'shake the bottle' must be taken seriously. Blood behaves thixotropically: on standing, red cells aggregate to form rouleaux. Changes of this type have been described in mucus. Gastropods move using a single appendage, the foot. Observations of a certain slug have shown that this mucus can change from being a glue, assisting adhesion, to being a liquid and so allowing for progression. The alterations are caused by a muscular wave sweeping forward on the foot (Denny 1980)."

Hill (1968) attributed the short-range stiffness he observed to the formation crossbridges between actin and myosin filaments of the muscles. This may be the basis of the thixotropic changes. It may be supposed that motion beyond a certain range tears the cross-bridges, which take time to reform once agitation ceases."

Walsh 1992, pp84-85.

Accordingly, the previous history of movement of a muscle affects its future performance when measured over short intervals. This phenomenon of the persistent effects of a muscle's previous movements has been referred to as a form of intramuscular "memory" (Lakie, Walsh and Wright, 1984a and 1984b). A common example of "thixotropy" is the stiffness experienced at the beginning of physical exercise and the looseness of movement experienced after "limbering up" which persists while movements continue, but is

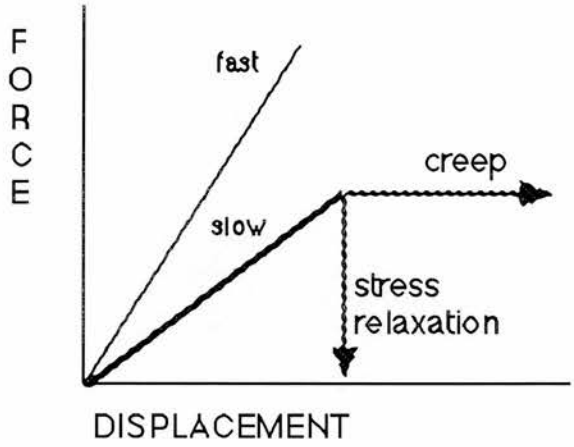
followed by stiffening of the muscles some time (but not immediately) after the movement ceases. Thus the unchanging posture, such as adopted by the author typing this thesis, results in stiffness during which the muscles adopt the characteristics of a 'gel' which in turn can be returned to a 'sol' state if the muscles are 'stirred' ie the author gets up and stretches.

Abrupt stretching or 'stirring', to use the Péterfi (1927) vocabulary, applied to passive and active muscles has been used extensively in experiments on human muscle thixotropy in awake and anaesthetised adults and at different bias angles at most of the major upper and lower limb joints of the body (Lakie, Walsh and Wright, 1984a and 1984b, Walsh 1992). The concept of thixotropy, or that the behaviour of muscle changes with handling, is likely to become increasingly important because of its implications for therapy and for studying muscle neurophysiology. For instance, thixotropic properties of the muscle spindle have recently been described (Hagbarth *et al*, 1987), indicating that even electrically-dependent events are altered by the physical properties and behaviour of muscle. Such experiments demonstrate that 'physiotherapy,' or the physical handling of muscles, at least temporarily, has a marked influence on neurophysiological response of such muscles in both health and disease. Thixotropy can be studied using the same apparatus as that used to assess elasticity, viscosity and inertia, under appropriate conditions (see Walsh, 1992 pp78-102 for a review).

Two other plastic features of muscle are recognised, 'stress relaxation' and 'creep' and are illustrated schematically in figure 2.2.6.2, above, and discussed below.

2.2.6 2. Stress relaxation.

"Stress relaxation" is observed if a resting muscle is stretched to a preset length, and maintained at that length: then the force (torque acting at a joint) required to maintain that length declines over time (Wright and Johns, 1960, and Foley 1961).The principle of stress relaxation is undoubtedly used daily in households with children and adults whose muscles require splints to maintain muscle stretches in order to preserve a functional angle such as the right angle at the ankle (also referred to as the anatomical, the neutral or 'zero' angle when the foot is neither plantarflexed nor dorsiflexed). After the initial slow morning stretches to the calf muscles, often requiring considerable applied torques, the ankle achieves neutral and a splint can be worn comfortably all day with minimal discomfort, presumably because the intramuscular tensions are very low, in contrast to a dystonic or 'electrically active' limb in



Plasticity or visco-elastic stiffness:
The elastic stiffness of a plastic substance is greater the faster the speed of stretch (steeper slope). Maintaining a constant stretch force results in continued elongation (creep). Maintaining a constant length causes the force of stretch to wane (stress relaxation).

Figure 2.2.6.2 Plastic properties of muscle. *after Wright and John's, 1960.*

which tension is likely to remain high throughout the day, with a strong likelihood of unwanted pressure sores as a consequence. (see section 3.2)

2.2.6.3. Creep.

"Creep" is observed when resting muscle is stretched with a preset but constant force: there is an initial rapid increase in length, but if the force is maintained, the muscle gradually lengthens further, albeit at a slower rate (Wright and Johns 1960). This has to be distinguished from electrically active eccentric muscle contractions opposing muscle lengthening or reflex resistance to stretch. Under such circumstances, the muscle behaves like a stiff putty, capable of remaining at its new length with little or no additional stretch force. When a muscle exhibiting creep is released it slowly returns to a longer resting length than at the start of stretch, equivalent to the original resting length plus the length achieved during the creep phase; slowly returning to the original length after many seconds or minutes (Wright and Johns, 1960). This slow return back to a baseline length was also recognised by Herman (1970) as an end-stage in the evolution of acquired hemiplegia in adults who had suffered strokes and in whom there was little (if any) residual evidence of reflex excitability: Herman's group IV hemiplegia. Herman (1970) viewed this state of the muscles as a pre-contracture stage.

2.2.7 Muscle Contracture.

The pathologically short muscle or tendon will offer a reduced joint range when compared to normal ranges. Attempts to stretch such muscles beyond their restricted range results in a rapid rise in tension, as when loading a spring. It is a feature of muscle, that sarcomeres will be added or removed if held in a lengthened or shortened state respectively.

"When the soleus muscle is immobilised by plaster cast in a shortened position, the muscle fibers lose 40% of their sarcomeres (Tabary *et al* ,1972). The immobilisation of this muscle at its maximum length results in production of 19% more sarcomeres. These changes take place within four weeks. They result in a change of the tension-extension curve, which depends only on sarcomere number (Tabary *et al*, 1976; Williams and Goldspink, 1978). All these changes are quite reversible. When the muscle is immobilised first in the shortened position and then in the more lengthened position, sarcomere numbers return towards normal despite doubling of the immobilisation period."

"Similar changes are observed in the length of the belly connective tissue, as demonstrated by the shift in and increased steepness of the passive curve (Tardieu *et al* , 1982). No excessive connective tissue is observed histologically. There is even an absolute decrease."

"Such an adaptation of the muscle structures is observed during normal growth, that is, without any experimental intervention. For a given joint angle, the sarcomere length remains the same at all ages, though the bone length and the sarcomere number change

considerably, being much greater in older animals (Tardieu *et al*, 1977). This shows that in young, normal animals muscle growth is closely related to bone growth, indicating the existence of a trophic regulatory effect."

Guy and Catherine Tardieu, 1987.

The importance of an appropriate and functional muscle length occupies a significant place in the experimental work of this thesis (see section 3). Tardieu and Tardieu (1987) highlight the differences in response to surgical tendon lengthening depending on the clinical phenotype of the child being operated upon. A restoration of the normal functional joint angle and torque generation is seen after lengthening in children with cerebral palsy, provided attention is paid to the mechanisms of abnormal muscle contractions attempting to return the muscle to its shortened length: the additional treatments advised include, where appropriate, braces; anxiolytic drugs; amphetamines; 45% alcohol injections to relieve increased stretch reflexes or cocontractions; nerve crushes or intramuscular 96% alcohol injections. In a second group of children with cerebral palsy without excessive motor activity, elongation corrects the joint angle but active force generation never returns to normal at the appropriate functional angles: the muscles remain permanently weak, although the contracture is unlikely to recur.

2.3 Neurophysiological Components of Muscle Tone.

2.3.1 Normal Muscle Tone.

Clearly, in addition to the underlying structural properties of muscles, tendons and joints, the appreciation of clinical tone in health and disease may be altered by a variety of mechanisms which, when disordered result in abnormal (reduced or increased) muscle tone. This has led to a number of studies summarised by Burke and Gandevia (1993):

" By analogy with the stretch reflex of the decerebrate cat, it is widely held that normal muscle tone is dependent on the 'tonic stretch reflex', and for this reason the description in 1966 that muscle vibration could evoke a tonic reflex contraction in both cat (Matthews, 1966) and man (de Gail, Lance and Neilson, 1966; Hagbarth and Eklund, 1966) met with wide interest. Hitherto, only phasic reflexes such as the tendon jerk and the H reflex could be studied in human subjects: a tonic reflex contraction in response to muscle stretch is not normally elicitable unless the subject performs a reinforcement manoeuvre or deliberately contracts the muscle being stretched (Neilson this volume). This elusiveness of the tonic stretch reflex in relaxed subjects raises the question whether reflex mechanisms normally contribute to muscle tone."

"The characteristics of the tonic vibration reflex (TVR) support the view that stretch reflex mechanisms have little to do with normal muscle tone. Tendon vibration can create an intense afferent barrage, predominantly though not exclusively in group 1a afferents, sustained for as long as the vibration is sustained (Burke *et al*, 1976a) unless a TVR develops and unloads muscle spindles (Burke *et al* ,1976b). The intensity of the afferent input exceeds that when non-contracting muscles are passively stretched. Despite the intensity of

the input to the spinal cord, the TVR develops slowly if at all, generates little force and is readily suppressed by the subject (de Gail et al 1966; Hagbarth and Eklund, 1966). Thus in relaxed subjects, the 'gain' of the reflex pathways is initially quite low, though it then increases slowly. By contrast, if the excitability of the reflex pathways is enhanced by asking the subject to perform a voluntary contraction or if the subject suffers from spasticity, the reflex response to vibration appears more rapidly and and reaches a plateau more quickly (Hagbarth and Eklund, 1968; Burke Andrews and Lance, 1972). These characteristics suggest that the reflex mechanisms are quite sluggish in relaxed man and can make little contribution to muscle tone. Accordingly, limb stiffness is not altered when neurologically normal subjects are rendered flaccid by general anaesthesia, in order to remove neural contributions to tone (Lakie et al 1980) and, as all electromyographers can confirm, resting muscles are electrically silent unless the patient tenses them."

"What then determines normal tone? To a clinician, muscle tone is the resistance to stretch felt by an examiner when he passively moves a joint of a patient to be relaxed. some subjects find it impossible to relax completely or will insist that they are relaxed when they are clearly not, particularly with proximal muscles. When a subject is fully relaxed, muscles are EMG-silent, and all but the most abrupt passive movements fail to evoke a reflex contraction. In such subjects, any resistance to stretch is due to passive viscoelastic and (thixotropic) properties of muscle, non-contractile tissues and joints (Lakie, Walsh and Wright, 1984*). The stretch reflex then makes no contribution to 'tone', whatever the level of fusimotor drive. In patients who fail to relax, passive stretch can evoke a reflex contraction , and in these patients the resistance to stretch will be determined by a combination of non-neural factors, the unintentional 'voluntary' contraction and the evoked reflex activity. In these patients, fusimotor drive could contribute to muscle tone. However, underactivity of the fusimotor system would not be detected because, at most, it could only remove something that is not effective in fully relaxed subjects with normal muscle tone."

Burke and Gandevia 1993, pp97-99

* reference 1984b in this text.

2.3.2 Hypotonia.

Surprisingly little is known about the mechanism(s) of hypotonia which represents a a state of reduced resistance to stretch and slackness of the muscles characterised in infants by:

"1. bizarre or unusual postures
2. diminished resistance of the joints to passive movement
3. increase in the range of movement of the joints."

Dubowitz 1980, p1.

Hypotonia may arise at any time of life through a variety of congenital or acquired lesions or may be normal at different stages of maturation of the foetus or infant:

"Muscular hypotonia is a common symptom in infancy. It may be the presenting feature of a neuromuscular disorder; it may occur in mentally retarded children or in the early phase of cerebral palsy; it may be a manifestation of a connective tissue disorder; it may be associated with various metabolic disorders in infancy; it may be an incidental and non-specific sign in any acutely ill child; it may be completely physiological in the premature infant; and it may occur as a completely isolated symptom in an otherwise normal child."

Dubowitz 1980, p 1.

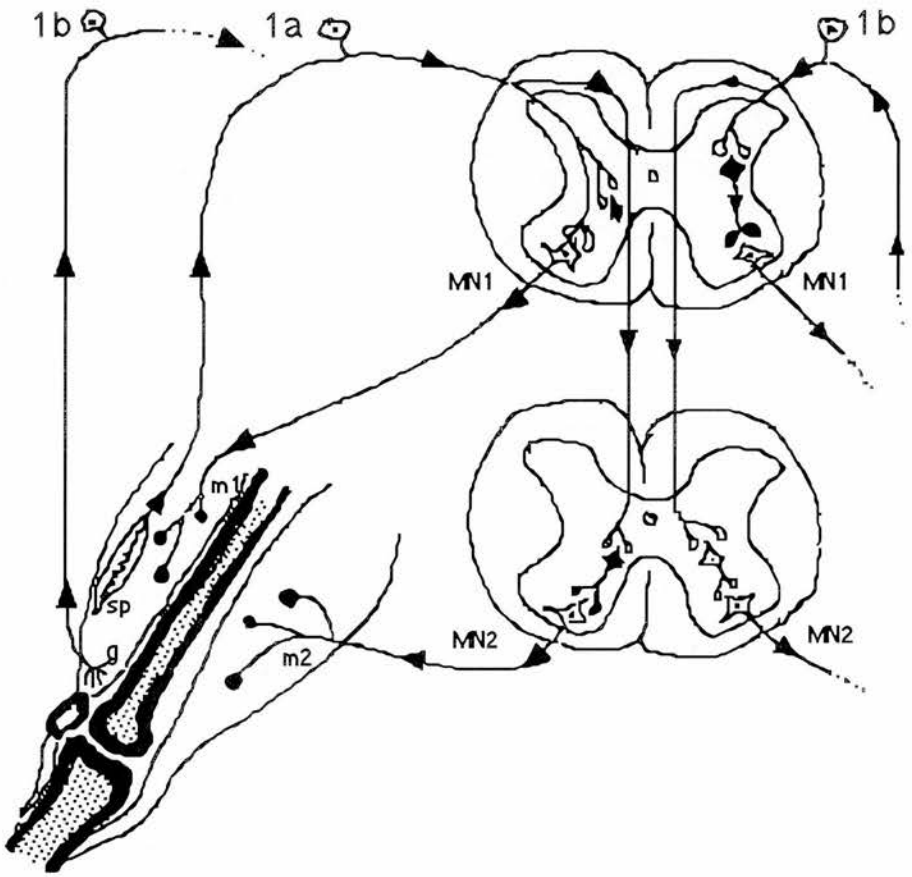


Figure 2.3 Diagram showing the stretch reflex pathways. Impulses from the muscle spindle (sp), in parallel with the extrafusal fibres of the muscle, travel up the large diameter 1a fibres to synapse with the ipsilateral motorneurone (mn1) which sends fibres back to the muscle via endplates (m1). The 1a fibres also connect with inhibitory interneurones (black structures) supplying ipsilateral antagonist motorneurones (mn2). Golgi tendon organs (g) located in the musculo-tendinous regions of the muscle, are stimulated by muscle contraction (as opposed to passive stretch) and send impulses up the 1b large diameter fibres and synapse with inhibitory interneurones (black structures) which in turn synapse with ipsilateral motorneurones (mn1). The 1b fibres send collaterals to excitatory interneurones (open structures) supplying ipsilateral antagonist motorneurones (mn2).

redrawn from Brodal 1981 & Eccles 1957

Burke and Gandevia (1993) have summarised the principal clinico-pathological features of non-neuromuscular causes of hypotonia:

" Hypotonia is said to occur in lower motor neurone lesions, in cerebellar disease and in spinal or cerebral shock. In none of these changes is fusimotor underactivity responsible for the 'disordered tone'."

"The reflex arc can be interrupted in lower motor neurone lesions, but hypoactive tendon jerks are found particularly when there is also involvement of the afferent side of the reflex arc. A peripheral lesion that affects only motor units cannot abolish tendon jerks unless it involves all low-threshold reflexly accessible motorneurones. If the motor involvement is random (or preferentially affects large motorneurones and large axons, as compressive lesions do), absence of the tendon jerk in a muscle that can still contract betokens a sensory disturbance. However, even when the tendon jerk is depressed, a decreased resistance to stretch is not due to absent reflex activity but to soft tissue change."

"In *cerebellar disorders* and *shock syndromes*, the low level of muscle tone is accompanied by other evidence of abnormality: dysmetria, dysdiadochokinesis, intention tremor and rebound in cerebellar disorders; flaccidity with tendon jerk areflexia, with or without loss of cutaneous reflexes in shock syndromes. By itself, and in the absence of collateral signs, muscle tone is unremarkable, not different to that seen in normal subjects who relax fully (Van der Meché and Gjin, 1986). In chronic cerebellar disturbances, soft tissue changes can occur, leading to enhanced compliance of muscle and joint, such that joints in a relaxed limb can adopt abnormally lax positions (see Lance and McLeod, 1981). Presumably such changes occur because the patients have lost the 'shock absorber' action of the stretch reflex during voluntary activities, and accordingly the hypotonia at rest could be attributed to an indirect effect of diminished reflex responsiveness."

Burke and Gandevia 1993, pp99-100:(authors' italics)

Another operational definition of hypotonia is the inability to compensate for external perturbations. Normally, a variety of physiological processes protect the body from such perturbations (Struppler *et al*,1987) and these can be classed according to their respective latencies and according to an anatomical hierarchy of structures effecting the resistance:

1. Elasticity and contractility of activated muscle which counteracts external disturbance.
2. Proprioceptive reflexes: spinal (short latency -oligosynaptic) or supraspinal (medium to long latency - polysynaptic and/or trans- cortical).
3. Voluntary compensation.

(*after Struppler et al 1987, see also fig 2.3.2*)

Table 2.3.2.1 lists the anatomical sites commonly associated with hypotonic states (excluding neuromuscular causes):

Table 2.3.2.1 Different causes of hypotonia and their influence on the phasic stretch reflex.

Site of lesion	Altered physiological process	Phasic stretch reflex
Dorsal spinal roots	spindle afferent traffic reduced	reduced
Differential nerve block	gamma efferent traffic reduced	reduced
Dorsal column	ascending spinal afferents reduced	normal
Cerebellar	alpha-gamma linkage reduced	normal
Thalamotomy or subthalamotomy	ascending muscle afferents ?	normal

after Struppler, Riescher and Gerilovsky 1987.

Hypotonia has been investigated with some interesting studies on adult subjects who had undergone stereotactic ablation of the nucleus ventrolateralis (VL) of the subthalamus (Struppler *et al,* 1987) to abolish resting, postural and intention tremor. This surgery also had the effect of decreasing rigidity, tonic reflex excitability and the effect of Jendrassik's manoeuvre (mental drive) on the reflex excitability and rigidity. Ventrolateral thalamotomy also diminished the rebound phenomenon following sudden unloading of an isometrically activated muscle in a limb supporting a weight, but force development, rapid movements and phasic stretch reflexes remained unchanged (Table 2.3.2.2)

Table 2.3.2.2 Clinical features of thalamotomy and subthalamotomy for Parkinsonian rigidity and tremor.

Variable	Effect
rigidity	reduced
tremor	reduced
Tonic stretch reflexes	reduced
postural tone	reduced
load compensation	reduced
phasic stretch reflexes	unchanged
maximal force generation	unchanged
muscle sense (force and movement perception)	probably unchanged

after Struppler et al 1987

By comparing the effects of selective gamma fibre xylocaine nerve block on the elbow flexor responses to perturbations with responses in subjects who had undergone very selective thalamotomy in the treatment of parkinsonian rigidity and tremor, Struppler and colleagues were able to identify two load resisting mechanisms: **a.** early compensation of sudden external disturbances before reflex onset, and **b.** compensation via the reflex arc (fig. 2.3.2).

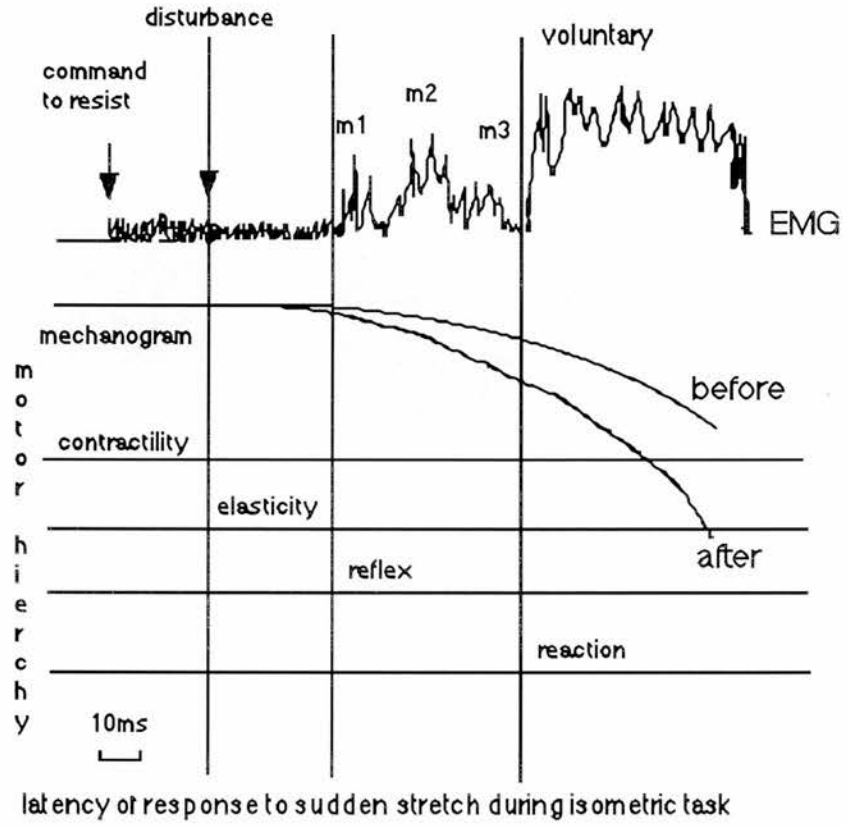


Figure 2.3.2. Mechanisms of resistance to muscular perturbation during an isometric task. Before and after stereotactic thalamotomy / subthalamotomy to abolish tremor and rigidity in Parkinsonism. After the command to "resist" with an isometric contraction, the muscle is suddenly stretched "disturbance". In the post-stereotactic patients the torque-induced elongation of the muscles is increased: the muscle appears to have reduced intrinsic stiffness. After stereotactic surgery, the mechanogram shows a reduced resistance even before the onset of the reflex responses. EMG=electromyogram, m1=short latency or spinal reflex, m2=polysynaptic spinal reflex, m3=long latency or transcortical reflex. Horizontal lines: ascending hierarchy of motor structures involved. Vertical lines: latency of recruitment of motor structures in ascending order from muscle to cortex.

Redrawn and modified after Struppler et al 1987

2.3.3 Myotonia and related conditions.

This is a failure of the muscles to relax or sustained contraction which is accompanied by an excessive electrical discharge causing the audio signal of the electromyogram to sound like a dive bomber (in the pre jet age!). Myotonia is neither a feature of cerebral palsy nor late-acquired or adult upper motor neurone syndromes, but is usually classed as a neuromuscular disorder (for a review see Dubowitz 1995). The myotonias are included in this classification of resistance to stretch purely for comparison with passive, intramuscular, biomechanical and 'extramuscular' causes of active muscle contraction. The pathophysiology of myotonia is attributed to a disturbance in ionic transport which alters the threshold for muscle depolarisation owing to a conductance change rendering it closer to the firing threshold. Compared with the well-known dominantly inherited dystrophia myotonica and myotonia congenita, rarer forms such as chondrodystrophic myotonia (Schwartz-Jampel syndrome) are accompanied by continuous electrical muscle activity which is difficult to relieve.

2.3.4 Reflex excitability and spasticity.

As already discussed in the sections on normal tone and hypotonia, the classical view, elaborated by Sherrington, of the stretch reflexes as a 'fractional manifestation' of the factors sustaining muscle tone has undergone extensive investigation over the years.

Brodal (1981, p163) summarises the principle components of the reflex arc as comprising a *receptor*, an *afferent conductor* having its cell body in the spinal or cranial nerve ganglia, a *reflex centre* where afferent messages and signals from other sources are processed, an *efferent conductor* going to the *effector* organ and an effector which produces the reaction (see fig. 2.3 above). In addition to the stretch reflexes (see below) more complex reflexes have been observed and described:

> "Many types of reflexes may be distinguished. Flexor reflexes are those in which the response is a flexion of the limbs. The most potent stimuli are nociceptive ones, and the result is a withdrawal of the limb (withdrawal reflex). In other reflexes there is an extension of a limb; for example, in the crossed extensor reflex which may accompany a flexor reflex. There are still other more complex reflexes; for example the scratch reflex. All of these reflexes usually involve several muscles, and the reflex response may vary according to the situation (type and site of application of the stimulus, concomitant application of other stimuli etc.) the arcs for these reflexes are of great complexity. Quite another type of reflex is the *stretch reflex, the contraction of a single muscle in response its being stretched.* This is the elementary reflex which probably occurs in all muscles."
>
> *Brodal 1981. p 163. (author's italics)*

Stretch reflexes may be classed according to:

1. latency of onset: short, intermediate or long.

2. mode of stimulation:length-dependent (tonic) or velocity- sensitive (phasic)

or in engineering terms according to the phase angle of the stimulus.

3. stereotypy: this relates to clinical judgments about normal or abnormal responses.

Stretch reflexes may also be quantified according to:

1. velocity threshold of excitation

2. velocity gain.

A history of the subject now requires an understanding of the concepts of 'the motor unit', 'fibre type' and 'motor unit recruitment threshold', the orderly recruitment of motor units now known as the 'Henneman Principle': in other words, a synthesis of basic interactions of muscles, nerves and the central nervous system (CNS).

2.3.4.1 Stretch reflex latencies.

The simplest stretch reflex , used clinically as a test of functioning electrical circuitry of the reflex arc has long been considered to be monosynaptic, although this is now disputed (see Burke and Gandevia, 1993, and discussion below). It has been observed within 25ms of the stretch stimulus for the elbow flexors and within 35 ms for the calf muscles in previously resting muscles. If the muscles are voluntarily (or involuntarily) active, a later electromyographic (EMG) discharge may be observed which is too soon to constitute a voluntary reaction: so called intermediate and longer latency reflexes thought to be mediated by spinal polysynaptic pathways (intermediate or M2 reflex) and transcortical pathways (M3 reflex), see figure 2.3.4.3 below (after Lee and Tatton, 1978).

"When discussing the stretch reflexes and their anatomical basis, it is appropriate to start with a relatively simple model, that of the patellar reflex, routinely tested in every clinical neurological examination. This is the *monosynaptic* reflex, the single synapse being in the spinal cord. It should be pointed out that, however useful clinically, this reflex probably has little or no functional significance, the necessary stimulus being highly unphysiological. Furthermore the reflex does not occur in all muscles, and is entirely absent in some healthy subjects. Reflex responses to stretch of a muscle, occurring with a longer latency than the monosynaptic stretch reflex, are apparently of greater functional significance and have received increasing attention since first described by Hammond (1956). Less is known about the pathways involved in these polysynaptic stretch reflexes."

Brodal 1981, p163

The functional importance of the short latency reflex has repeatedly been called into question in relation to all but the most trivial of tasks. It serves as some measure of intact connections in clinical neurology, but what does it actually tell us about the integrity of the motor system? In fact, much of what we know about the muscle spindles, sensory 1a afferents, descending supraspinal influences, the physiology of the motor unit and mechanisms of motor unit recruitment and alpha-motorneurone firing properties, both voluntary and reflex, have been gleaned directly or indirectly through the study of the behaviour of this simplest of reflex arcs in health and disease, using a variety of mechanical (tendon jerks, ramp and sinusoidal stretches; vibration) and electrical (H reflexes) stimuli.

Some of the assumptions underlying this simplest of motor pathways have been reviewed by Burke (1983) and Burke and Gandevia (1993).

"One of the few objective signs in clinical neurology is the tendon jerk. It is usually thought that an abrupt percussion on the appropriate tendon (i) directly stretches the relevant muscle, (ii) activates primary spindle endings in that muscle, and (iii) produces a synchronised volley in group Ia afferents from the muscle, and that (iv) this afferent input also produces a tendon jerk through a monosynaptic pathway. It was once taught that, (v) in the absence of background fusimotor drive, spindle endings would be insufficiently sensitive for tendon percussion to evoke a reflex response. None of these views can be accepted without qualification."

"(**i**) percussion-induced direct muscle stretch is not necessary to evoke a tendon jerk (Lance and de Gail, 1965). Indeed, when percussion is delivered so that it shortens the relevant muscle and tendon, a tendon jerk still occurs. In subjects with hyperactive tendon jerks, percussion on a nearby bony prominence is often sufficient to evoke the reflex contraction, and in patients with pathological hyperreflexia, percussion on the clavicle can produce reflex contractions in upper-limb muscles, and percussion on the pubis can produce reflex contractions in lower limb muscles, particularly the adductors of the thigh. 'Irradiation of reflexes' in patients with spasticity is due not to intraspinal spread of an afferent volley from the percussed muscle but to spread of the percussion-induced vibration wave through bone to spindle endings in remote muscles (Lance and de Gail, 1965). In other words, the reflex contractions of muscles remote from the percussion site occurs because the percussion wave stimulates spindle endings in these remote muscles. Direct support for these conclusions came from experiments in which afferent volleys were recorded using microneurography (Burke, Gandevia and McKeon, 1983), as described below."

"(**ii**) In the cat, abrupt muscle stretch can be applied directly through a tendon in such a way that it activates selectively primary spindle endings in the stretched muscle (Lundberg and Winsbury, 1960); Stuart, Willis and Reinking, 1971). However, tendon percussion involves a transversely applied indirect stimulus that is difficult to quantify let alone control. Percussion on the Achilles tendon is an effective stimulus for primary spindle endings in triceps surae, and responsive endings may discharge multiple times in any one percussion-induced volley. Activation of different spindle endings depends on transmission of the percussion wave to spindles dispersed throughout the muscle belly, and therefore does not occur at precisely the same moment for each ending. As a result, the group Ia volley from spindles in triceps surae is quite dispersed even at the popliteal-fossa level, taking some 10ms to reach its peak and lasting some 30-40ms (Burke *et al*, 1983). Tendon organs and secondary spindle endings may also respond to percussion, the former usually with only a

single discharge, the latter commonly as a transient increase in background discharge frequency. In addition the total afferent input produced by tendon percussion will involve group 1a activity from other calf muscles and even from pretibial and peroneal muscles, innervated by the peroneal nerve, Percussion insufficiently strong to produce an ankle jerk activates muscle and cutaneous afferents from the foot (Burke et al, 1983). Afferent volleys from this source take a few milliseconds longer to reach the popliteal fossa, but they can still contribute to the rising phase of the percussion-induced afferent volley. It can be concluded that the tendon percussion is not a clean stimulus: it is virtually impossible to produce a volley only in group Ia afferent s from the percussed muscle, especially if the intensity of percussion is sufficient to evoke an ankle jerk."

"(**iii**) As discussed above, the percussion-evoked group 1a volley from triceps surae is highly dispersed at popliteal-fossa level (Burke et al, 1983). In the human thigh, the conduction velocity of fast group I muscle afferents is approximately 60m/s (Macefield, Gandevia and Burke, 1989). If the slowest group Ia afferents travel at 40m/s, even a pure Ia volley evoked electrically by stimulation of the tibial nerve in the popliteal fossa would have a dispersion of approximately 5ms by the time it reached the motorneurone pool. A percussion-evoked volley will be much more desynchronised because it is already dispersed at popliteal-fossa level."

" (**iv**) The monosynaptic pathway would be the sole spinal pathway involved in the tendon jerk, only if other inputs failed to reach the motorneurone pool before the target motor neurones discharge. This condition cannot be fulfilled. Using two separate techniques (H reflexes conditioned by subthreshold tendon percussion; and poststimulus time histograms of voluntary active motor units in soleus in response to tendon percussion), it has been found that the rise-time of the compound excitatory postsynaptic potential (EPSP) produced by tendon percussion lasts >10ms, even when the intensity of percussion is too weak to evoke an ankle jerk (Burke, Gandevia and McKeon, 1984). Motorneurones which are only just recruited into the reflex discharge will be activated some 10ms after the onset of monosynaptic excitation. There is adequate time for interneuronal transmission from homonymous and antagonist Ia afferents and from Ib afferents to reach late-recruited motorneurones before they discharge. In addition, 10ms is sufficient for recurrent inhibition from low-threshold motorneurones to affect the discharge of higher threshold motorneurones, and for the earliest group I impulses to evoke presynaptic inhibition of slow Ia afferents or of the second impulses in fast Ia afferents. The situation may be even more complex. The only certainty is that pathways other than the monosynaptic pathway will be active, and that these pathways will affect the recruitment of some motor neurones."

"In a strong reflex contraction, the first recruited motorneurones may discharge early on the rising phase of the compound EPSP, and so might be activated exclusively by monosynaptic inputs. However in relaxed subjects, much of the compound EPSP must be spent raising the motorneurone pool to firing threshold (Burke *et al* ,1984), so that even with large rapidly rising EPSP's there will be some delay before low-threshold motorneurones reach threshold. Moreover, an increase in tendon jerk amplitude is produced by recruiting motorneurones that had previously not been recruited; inhibition of the tendon jerk will affect preferentially those motor neurones that had only just been recruited. In both cases, this will occur at the peak of the compound EPSP > 10ms after its monosynaptic onset. It is of interest that oligosynaptic Ia excitation of homonymous and heteronymous motorneurones has been documented for lower-and upper-limb muscles of human subjects (Fournier *et al*, 1986, Malmgren and Pierrot-Deseilligny, 1988)."

"(**v**) Tendon jerks can be elicited in relaxed normal subjects. There is considerable evidence that the level of fusimotor drive directed to spindle endings in relaxed human muscles is quite low, possibly negligible (Vallbo et al 1979; Burke, 1981). Recently, evidence has been found for background fusimotor activity directed to spindle endings in EMG-silent pretibial muscles of subjects who were standing upright (Ribolt, Roll and Vedel, 1986; Aniss

et al 1990), but this does not alter the generality that, for muscles not engaged in any task, there is little background fusimotor drive. This view is supported by nerve block experiments, in which the percussion-evoked volley from soleus was recorded from the tibial nerve in the popliteal fossa while the sciatic nerve was blocked completely by concentrated lignocaine (Burke, McKeon and Skuse 1981b). There was no change in the evoked afferent volley when all fusimotor influences were eliminated. The tendon jerk can be potentiated by a number of different manoeuvres including voluntary contraction of a remote muscle, contraction of the test muscle, discomfort, arousal and a decrease in percussion rate. None of these manoeuvres enhances the muscle afferent response to percussion (Burke et al 1981a). This indicates that changes in amplitude of the tendon jerk may be produced centrally by altering reflex transmission within the spinal cord, rather than peripherally by altering the muscle afferent input to the spinal cord. Similarly, in spasticity, the site of the reflex enhancement appears to be central rather than peripheral. "

Burke and Gandevia 1993, pp91-97.

This somewhat exhaustive citation by Burke and Gandevia serves to enumerate and answer some of the fundamental issues in relation to the anatomy and physiology of the tendon jerk. Most of the foregoing research has focussed on the stimuli facilitating or inhibiting the tendon jerk, relying on microneurographic studies of impulses generated in the Ia afferents. The concepts of conditioning stimulus, adequate stimulus, dispersion, presynaptic inhibition and modulation, motorneurone excitation threshold and recruitment, will be of material importance to the understanding of some of the experimental work performed for this thesis.

2.3.4.3 The Hoffmann reflex.

Considered to be an electrical counterpart of the mechanically-induced tendon jerk, the Hoffmann reflex (1922) is obtained by electrically stimulating the afferent nerves, principally the group Ia afferents, which in turn causes motorneurone depolarisation and a reflex twitch. If the EMG is recorded, an initial EMG discharge and muscle twitch corresponds to direct motor axon stimulation which produces muscle depolarisation (M response) followed some 30-35 ms later by a second, reflex EMG discharge (H-response, relayed via the spinal cord) and a second muscle contraction of the soleus muscle. Early studies have included those of Magladery *et al* (1952) and Paillard (1955) whose findings are discussed in relation to some of the experimental results below (section 3.3.1). Theoretically at last, here was a technique which could be used to test the integrity of the spinal reflex circuitry with reproducible stimuli of known current or voltage, while by-passing the muscle spindle and its mechanical intricacies. Much of the ensuing work has concentrated on providing standardised methods of elicitation for use in studying disordered motor states (Hugon 1973, see section 3.3.1 below), particularly with respect to patient and electrode positioning,

stimulus duration and intensity and the interpretation of the so-called 'recruitment curves' of the M-and H-responses in relation to stimulus intensity. One of the aims of such studies was that they might aid in the quantitation of 'spasticity', since in disorders of the upper motor neurone, it was noted that the short latency reflexes were of larger amplitude, and more easily elicited than in healthy individuals. The method involves expressing the H-reflex amplitude as a fraction of the M response on the assumption that the M response is constant in health and disease, but the H reflex amplitude is increased with spinal or cerebral corticospinal damage (see 'spasticity' below).

Quite clearly, the H-reflex is truly unphysiological, being the product of a wholly artificial means of nerve stimulation. Even the humble tendon jerk has a greater claim to correspondence with a physiologically plausible stimulus as might be encountered when landing on ones toes such as in sprinting or jumping, nevertheless, tendon jerks, ramp and sinusoidal muscle stretches at different velocities and frequencies respectively along with the H-reflex testing provide interesting insights to motor behaviour.

Once again, Burke (1983) highlights some essential distinctions to be borne in mind when attempting to compare the tendon jerk and H-reflex:

> "In clinical neurophysiology, it has become almost axiomatic that the tendon jerk and the H-reflex are essentially identical reflexes which differ only in that the tendon jerk requires the integrity of the muscle spindle, and that differences in the behaviour of the two reflexes accurately reflect the level of fusimotor drive. At its best, this is an oversimplification. In spastic patients, it is commonly held that the tendon jerk is exaggerated more than the H-reflex. The discrepancy in the degree of accentuation of these reflexes has been taken as evidence that fusimotor tone is elevated in spasticity. This conclusion assumes that (a) an increase in fusimotor drive would produce a parallel increase in the tendon jerk; (b) the level of fusimotor drive is the only significant factor affecting muscle spindle responsiveness; and (c) the only significant difference between the tendon jerk and the H-reflex is the contribution of fusimotor activity in the former."
>
> *Burke 1983, p137-138.*

Reviewing the literature available at the time, Burke (1983) is unable to find evidence for increased fusimotor drive that could explain a greater exaggeration of tendon jerk reflex response over that of the H reflex, except in the case of the voluntarily activated muscle. With regard to muscle spindle sensitivity, other factors might contribute to increase such sensitivity, particularly the disuse and atrophy of chronic immobility (Williams, 1980) or following steroid treatment (Botterman *et al*, 1981). A more subtle possible alteration of the spindle sensitivity could be due to the initial stiffness of the actin-myosin bonds of the intrafusal fibres (Brown *et al* 1969) resulting in a high frequency initial burst of Ia afferent

impulses at the onset of movement. Finally, the idea that background fusimotor activity only facilitates the tendon jerk is rejected on the grounds that the background spindle activity alters the excitability of the motor neurone (MN) pool, however it may be stimulated.

Aside from the disputed issue of background fusimotor drive, Burke (1983) discusses at least six differences between H reflexes and tendon jerks.

i) Distribution of the reflexes.

The H reflex is most easily elicited in the triceps surae, but in the upper limb, especially, requires some form of background excitation of the motor neurone pool, eg a weak contraction, the performance of the Jendrassik manoeuvre, the use of a pair of stimuli to elicit the reflex (ie an initial conditioning stimulus) or the presence of spasticity.

ii) The effect of ankle joint angle.

Rotation of the ankle joint affects the excitability of the soleus MN pool due to changes in the lengths of its agonist, synergists and antagonists; the activation of cutaneous and joint receptors. So that passive dorsiflexion of the ankle results in inhibition of the soleus H-reflex in normal man (Mark *et al*, 1968; Delwaide, 1971). If the muscle stretch produced by dorsiflexion altered the responsiveness of the primary spindle endings in soleus, percussion of the Achilles tendon could evoke a larger afferent volley. Provided the size of the afferent volley was sufficient to outweigh the inhibitory background activity, the triceps surae tendon jerk could be potentiated in the dorsiflexed position as reported by Herman (1969) within the range of 30° of plantarflexion to the zero position (shank at right angles to the sole of the foot). An apparent finding has been that in subjects exhibiting spasticity, the influence of stretch on the differences between tendon jerks and the H-reflexes is more pronounced: dorsiflexion facilitating the tendon jerk (Herman, 1969), but inhibiting the H-reflex (Burke *et al* 1971) in the same -30° to zero joint range:

> " The more dorsiflexed the ankle, the greater the the accentuation of the tendon jerk relative to the H-reflex. In most studies that have compared the the tendon jerk and the H-reflex in normal and spastic subjects, the angle of the ankle joint was not specified, and it is not always clear that the angle was the same for the tendon jerk and the H-reflex tests."
>
> Burke 1983, p139

iii) The afferent fibres activated by the stimuli.

H-reflex electrical stimuli activate Ia and Ib afferent fibres in roughly equal measure, though humans have been shown to have slightly faster Ia afferents which have slightly lower thresholds than Ib Golgi tendon organ afferents. (Pierrot-Deseilligny *et al*, 1981). Although

tendon taps can stimulate Golgi tendon organs, this is not as intense as primary spindle endings, so that an electrical stimulus, will activate proportionately more Ib afferents than tendon taps. As shown in figure 2.3., although the Ib pathway is disynaptic and afferent conduction in Ib fibres is slightly slower than in Ia fibres, there will be overlapping actions of Ia and Ib inputs to the MN pool following H reflex testing. Furthermore, the tendon tap percussion wave is bound to spread to antagonist pretibial muscles, whereas electrical stimulation of the tibial nerve will also excite the small muscles of the foot which are supplied by the posterior tibial nerve: whichever stimulus modality is used, contamination by afferent input from other muscles is likely. In the event of cutaneous afferents being stimulated, their slower conduction velocities preclude their input from reaching the MN pool during the composite group Ia excitatory post synaptic potential (EPSP).

iv) Discharge pattern.

A single afferent discharge occurs in Ia fibres following electrical stimulation, whereas, after tendon tapping, the spindle endings respond with bursts of impulses reaching frequencies of 100-200 impulses/s, causing repetitive discharges in individual Ia fibres, with intervals of 5-10ms or less between bursts, allowing for several bursts to influence the MN pool EPSP.

v) Dispersion.

As discussed above, with tendon percussion, the MN pool will have been conditioned only by fast conducting Ia afferents, but second discharges in fast-conducting Ia afferents may contribute to the reflex discharge.

vi) Motor neurones (MN) participating in the reflex contraction.

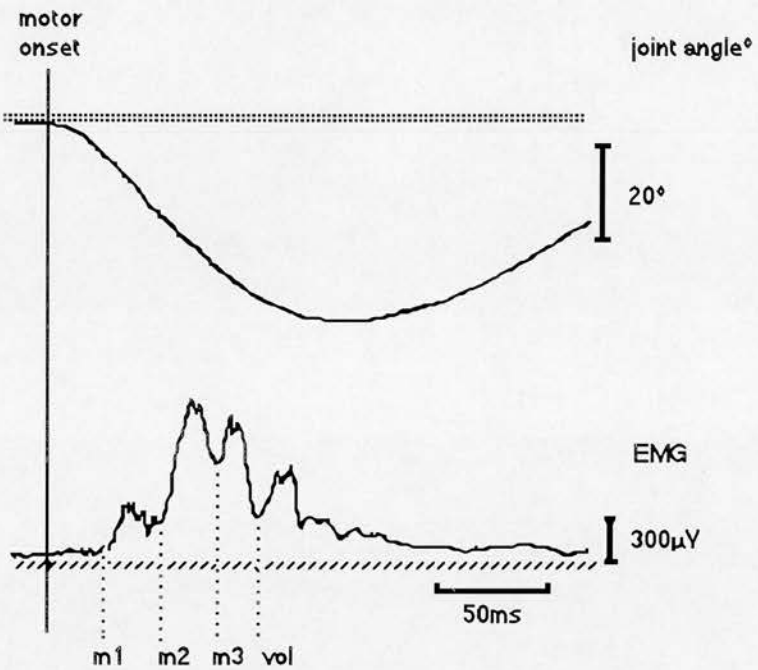
Perhaps the most significant difference between tap and electrically evoked reflexes lies in the populations of motor units which are excited.

"As usually elicited, the H reflex of triceps surae involves predominantly soleus motor units, although the M wave appears more readily in gastrocnemius (Homma and Kano, 1962; Levy, 1963; Buchtal and Schmalbruch, 1970; Hugon, 1973). On the other hand, the tendon jerk can be recorded in gastrocnemius as well as well as soleus (Homma and Kano, 1962), although it appears at lowest threshold in soleus (Levy, 1963). It is possible to record selectively from soleus using surface EMG electrodes distal to the insertion of gastrocnemius into the Achilles tendon (Hugon, 1973), but in this location a mechanical artifact may be recorded with tendon percussion. Hence, a more proximal and less sensitive site is likely to be chosen in comparative studies of the tendon jerk and H reflex potential."

"The basis of the low reflex threshold of soleus is probably that it is composed of low-threshold, slow-twitch motor units, whereas gastrocnemius contains both fast- and slow-twitch units, the former predominating in a ratio of 3:1 (see for example RE Burke, 1967, 1973). With low-intensity electrical stimulation, only small low-threshold motor units would be

activated reflexly. With stronger intensities, large motor units of higher threshold may well be activated reflexly but be unable to participate in the reflex EMG burst if, as is likely, their large-diameter axons were preferentially recruited in the M wave (Homma and Kano, 1962). It is common practice to use stimuli that produce a small M response, since if the M wave remains constant, experimentally induced changes in the H-reflex are probably not due to displacement of the stimulating electrodes. If there is an M wave, adjustment of stimulus levels so that the tendon and H reflex produce reflex EMG bursts of identical amplitude could result in the H-reflex stimulus activating a greater proportion of the MN pool than the tendon jerk stimulus. In addition, if there are differences in the MNs contributing to the reflex contraction in the tendon jerk and the H-reflex, the changes in motor unit properties that occur both in spasticity (Edström, 1970; Hopf et al, 1974; Mayer and Young, 1979) and as a result of inactivity (Davis and Montgomery, 1977) may well be a significant factor complicating the interpretation of comparative reflex studies."

Burke, 1983, p143.



Latency of stretch reflex responses following imposed flexion of wrist extensors.
m1=32ms, m2=59ms, m3=85ms, voluntary reaction=107ms.
after Lee and Tatton, 1978.

Figure 2.3.4.3 Long latency stretch reflexes.
The three stretch reflex responses, m1, m2, and m3 are elicited when a muscle exerting 10%-20% of maximum voluntary contraction is stretched: as can be seen, all three reflexes occur before the subject has time to react against the imposed stretch. The EMG is rectified.
redrawn after Lee and Tatton, 1978.

2.3.4.3 Long latency (transcortical) stretch reflexes in health and disease.

The elucidation of the physiology of the long latency reflexes has been lengthy and to some extent controversial as differing theories, with experimental observations to back

them up, have been advanced. The first requirement would be to demonstrate that afferent input from the muscle spindles reach the cerebral cortex and then, that the latency from the time of stretch to reflex EMG discharge was compatible with a transcortical route.

Oscarsson and Rosen (1963) demonstrated contralateral projection to cerebral cortex of muscle spindle afferent inputs along large diameter fibres from the forelimb of the cat. A few years later, Phillips, Powell and Wiesendanger (1971) demonstrated projection from low-threshold afferents of the hand and forearm to area 3a of the baboon's cerebral cortex, that is to say in a zone of the Rolandic fissure midway between the motor cortex anteriorly (precentral gyrus, area 4), and the sensory cortex posteriorly (post central gyrus, area 3).

Evidence to support the transcortical loop in humans followed in a series of publications, on latency experiments exploring the so-called servo mechanisms allowing the body to respond to perturbations more rapidly than would be allowed by voluntary reaction times in studies of the thumb (Marsden, Merton and Morton, 1972; 1973), and in other muscles (Marsden, Merton and Morton 1976). These studies being supported by observations of alterations in the firing patterns of corticomotorneurones in monkeys whose arms were perturbed while performing trained tasks (Evarts, 1973;Evarts and Tanji, 1975).

Long latency responses to stretch were studied in the muscles of the shoulder girdle and in particular, from a subject in whom the short latency monosynaptic stretch reflexes were absent or 'feeble' (Marsden, Merton and Morton, 1976) and the loss of such long latency responses in patients with cortical or internal capsular lesions (Marsden, Merton, Morton and Adam, 1977) lent further support to the notion that the long latency reflex was functionally present in intact man, but lost when the cortex or subcortical structures are damaged.

The ability to record the cortical evoked potentials from muscle stretches has been reported by Starr and colleagues (1981) after passive plantarflexion at the ankle in 10 healthy controls. The amplitudes of the cortical-evoked potentials were largest at the vertex and varied inversely with the repetition rate of the stretches. In a very elegant study (Abbruzzese *et al* ,1985), the cortical-evoked potentials from stretching wrist flexors in 9 healthy adult males were found to habituate (diminish) with increased rate of presentation of the stretch stimulus (ie at rates greater than 0.5 Hz) and the amplitude of the cortical potentials to increase with the velocity of stretch applied, demonstrating that the long latency reflexes are

velocity-dependent phenomena.

Despite the accumulating evidence for a transcortical anatomical pathway supporting the physiology of the long latency reflexes (see the volume devoted to the subject edited by Desmedt, 1978, and the extensive review by Wiesendanger and Miles, 1982), several alternative mechanisms have been postulated. One alternative hypothesis has been advanced by Eklund and colleagues who demonstrated intramuscular resonances of 30-50Hz set up within wrist muscles after brisk perturbations (Eklund *et al*, 1982a, b), such intramuscular oscillations increasing in amplitude with increasing speed of onset of movement and also after abrupt halts in stretching.

"It is concluded that segmentation of the reflex EMG responses to sudden joint displacements and other types of brisk muscle perturbations to a large extent depends on the inherent resonance characteristics of musculo-tendinous structures Primary spindle endings with their high vibration sensitivity and their segmental projections to alphamotor neurones are believed to be the receptors primarily responsible for reflex entrainment of the motor impulses."

Eklund et al 1982a

The latencies of the various reflexes in different muscles, usually ascribed to 'long loop' reflexes have been reinterpreted as possibly arising from intramuscular resonance:

"The interval between initial and second EMG peaks following tendon taps was longer for calf muscles than for wrist flexors and longer for wrist flexors than for jaw elevators. Similar differences were observed with respect to the intervals between the damped intramuscular oscillations initiated by the impacts.

"Without denying the existence of 'long loop reflexes' it is concluded that the characteristics of the late responses to muscle stretch in man are explicable also in terms of the resonance hypothesis."

Eklund et al 1982b

However, Matthews (1984) failed to demonstrate a significant long-latency reflex response following vibration at 143 Hz and in the later work, vindicating the transcortical route (1989) he writes:

"The vibration experiments may perhaps be telling us that the long-latency stretch response is highly discriminating in its afferent requirements and depends upon a specific pattern of afferent input, whether with regard to the types of fibre involved or the temporal sequencing of their discharges. Higher-level coordinating centres might demand a physiologically meaningful input and refuse to respond to the indiscriminate excitation of a single afferent channel, even though this suffices to elicit a short-latency response from spinal centres." *Matthews, 1989.*

Another possible alternative mechanism to support long latency responses has been advanced by Matthews (1984) and subsequently refuted by Matthews (1989). The earlier work had purported to show that the long-latency response could be transmitted by the small diameter, slowly conducting type II afferent fibres (Matthews, 1984), but after cooling experiments to the forearm, which abolished type II fibre conduction, the long latency response persisted (Matthews, 1989) indicating that the Ia afferent fibres were indeed mediating the reflex. Summing up the position, after years of studying alternatives to the trans-cortical route Matthews writes:

"The spinal stretch reflex exemplified by the tendon jerk appears to be less important in humans than a delayed 'long latency' response. This is easily observed when muscles of the hand are stretched while they are already contracting voluntarily. On limited evidence, many have long held that the delayed response is a transcortical reflex and have tended to neglect alternative possibilities, particularly that it might be a spinal reflex dependent upon slow afferents. New experiments have now eliminated the alternatives, leaving the transcortical hypothesis in command of the field.

Matthews, 1991.

2.3.4.4 Implications of the short and long latency reflexes.

The implications of the long-latency or segmental reflexes for motor control in man are that they appear to provide a mechanism for responding to external perturbations during voluntary and involuntary tasks at times when there is some background muscle activation. With cortical-subcortical damage, the evidence suggests that the pattern of long-latency responses is diminished or lost altogether by contrast to the short-latency responses which are enhanced or exaggerated. To some extent, the short-latency, or at least oligosynaptic, stretch reflex threshold, and gain appear to vary inversely with the longer latency reflexes. However the extent to which this reciprocal pattern obtains in different clinical cases is not entirely clear as in the cases described by Berardelli *et al* (1983) in which the long -latency reflexes were often preserved or enhanced:

"Electromyographic responses of triceps surae to dorsiflexion stretch were studied in 47 patients with a variety of lesions producing an upper motor neuron syndrome. The short latency spinal reflexes, both when the patient was at rest and when he was exerting a voluntary plantarflexion, were frequently enhanced in magnitude and the rate of increase with acceleration was also enhanced. Long-latency reflexes were uncommon at rest. With background force long-latency reflexes were present unless the short latency reflex was very large. Long-latency reflexes often were normal, but in some patients they were either excessively large or even of abnormal shape with prolonged continuous activity. The clinical assessment of the ankle jerk correlated with the magnitude of the short-latency reflex. The clinical assessment of tone correlated with the magnitude of the short-latency reflex, the magnitude of the long-latency reflex and the duration of the long-latency reflex. There appear to be multiple physiological mechanisms underlying the clinical phenomenon of spasticity."

Berardelli et al 1983

In the above study, 11 cases had cerebral palsy, 16 suffered from multiple sclerosis, 7 had had a stroke, 6 had cervical spondylosis, 2 had cortical tumours and 2 had motorneurone disease (amyotrophic later sclerosis). The last three cases had primary lateral sclerosis, chronic myelopathy and olivo-ponto-cerebellar degeneration respectively. In only 11 out of 47 cases were the long-latency reflexes abolished, compared to the 13 out of 14 cases with upper motor neurone lesions described by Marsden *et al* (1977). One explanation offered is that long-latency reflexes are lost acutely, which was when most of the Marsden *et al* (1977) patients were studied, whereas most of the Berardelli *et al* (1983) patients were in a chronic phase. A second possibility relates to the spectrum of anatomical sites of the lesions in the CNS between the two populations of patients.

A third difference relates to the physiology and pathophysiological differences between mechanisms affecting the thumb, as in Marsden's (1977) group and the triceps surae, which was the muscle of choice of the Berardelli (1983) group of patients. In this context, it has been postulated (Berardelli *et al* 1982) that the long -latency responses for the triceps surae depend on an *intraspinal long loop* as opposed to a *supraspinal long-loop* for the thumb muscles. This difference in the long-loop mechanisms of the thumb and calf muscles is supported by anatomical evidence from the cat in which forelimb (Oscarsson and Rosen, 1963) but not the hind-limb muscle stretches (Mountcastle *et al* 1952; McIntyre 1953, 1962) produced cortical potentials. These studies indicating the predominant cortical representation for hand and spinal representation for the leg muscles respectively. Unfortunately, the authors (Berardelli *et al*, 1983) do not describe any specific correlations of the short- or long-latency responses in their subjects with 'cerebral palsy': Nor do we know the nature of the CP phenotypes, except by inference from their selection of patients with 'upper motorneurone' lesions, that they were not 'dystonic' or 'athetoid' (see section 1.3.3 above: Classification of cerebral palsies).

2.3.4.5 Long-latency reflexes in extrapyramidal disease.

As a final note on the long-latency stretch reflexes, it should be borne in mind that they are enhanced in Parkinsonian states with background cog-wheel rigidity of muscles acting at the wrist (Lee and Tatton, 1975, Tatton and Lee, 1975, Lee and Tatton, 1978), at the thumb (Marsden *et al 1978*), the biceps muscle (Mortimer *et al* 1979) and tibialis anterior (Chan *et al*, 1979) in which the M2 and M3 long-latency reflexes are accentuated, being up to

five times as large as in normal subjects (Lee and Tatton, 1978, p 326). These patients do not need to activate their muscles with a mild contraction, as do normal subjects: consequently, the long-latency responses are elicitable even when the subjects are instructed to 'let go'. By contrast, Parkinsonian tremor alone without cog-wheeling, is not accompanied by an accentuated M2 or M3 response. One of the theories relating to this lack of modulation of the long-latency reflexes in Parkinsonism is that the corpus striatum usually sends projections to the premotor and motor cortex via the globus pallidus and thence to the ventrolateral thalamus (Lee and Tatton, 1978). It is speculated that the basal ganglia modulate feedback from the periphery, when this modulation is lost, the gain in the feedback loop may be constantly set at maximum (Lee and Tatton, 1978). Berardelli, Sabra and Hallett (1983) report similar findings for the triceps surae in Parkinsonian subjects and in addition, report a shortening reaction in the tibialis anterior muscles during ankle dorsiflexion. Nevertheless, Marsden *et al*, 1978, regard the enhancement of the long-latency response in Parkinsonism as a consequence of the increased background EMG as opposed to a true abnormality of the long-loop reflex pathways and their central modulation.

Thus, in contrast with upper motorneurone disorders in which the M2 and M3 responses may be diminished or abolished, they are consistently enhanced in Parkinsonism.

2.3.5 Spasticity.

2.3.5.1 What is spasticity?

As already mentioned in section 1 on the classification of the cerebral palsies, the term spasticity has been used freely and has permeated every aspect of the care for children with cerebral palsy: from establishing the epidemiology and natural history, to interpretation of the morbid pathology exhibited in early post-mortem studies (Christensen and Melchior, 1967) as well as inferences drawn from the latest neuroimaging techniques, and most importantly, to the selection of certain children for specific treatment (s). Yet in most instances, the term spasticity is used as a synonym for 'motor disorder' or 'cerebral palsy'.

There are several problems about this state of affairs, the first being that it implies that the major component of the motor disorder is related to abnormal tone and reflexes (as opposed to abnormal motor programming, posture, weakness and incoordination); that the spasticity interferes with the execution of voluntary movements; that relief of 'spasticity' would allow greater motor control to emerge and the detraction from developing other

strategies of care. These notions have been shaped by the history of clinical neurology, neuroanatomy and neurophysiology based on case studies and case series as well as attempts at establishing animal models of spasticity.

The risks of descending into a meaningless jargon has been highlighted by Landau (1974; 1988) who has questioned some of the underlying assumptions of loosely applied clinical terminology. Table 2.3.5.1 lists some of the commonly used definitions of spasticity discussed by Landau (1974):

Table 2.3.5.1. What is spasticity?

Spasticity defined as	Possible interpretations of the term 'spasticity'
1. Proprioceptive reflex release:	unstimulated muscle is quiescent, with increased phasic and tonic stretch reflexes and a clasp-knife response.
2. Generalised reflex release	including polysynaptic flexion reflexes eg chronic flexor spasms of chronic paraplegia.
3. Upper Motor Neurone Syndrome	poor motor performance and reflex release.
4. Dystonic-rigid state	ill-defined with many causes and pathological features, characterised by continuous muscle contraction: "busy line."
5. Mixed	Combinations of 1-4.
6. Undefined	

after Landau, 1974

Almost whichever of these possible interpretations are used, 'spasticity' remains a complex term with no clear definition, no clear link to a specific neuroanatomical locus or tract, (in spite of attempts to link it to lesions of the pyramidal tract, refuted by Tower, 1940) no clear implications for treatment and no clear indications that such treatment(s) would be of benefit to the patient. Landau (1974) was particularly anxious to separate out the different aspects of the motor syndrome as characterised by the early studies of Hughlings Jackson (see Taylor, 1958, *Selected Writings of John Hughlings Jackson*) in terms of the negative manifestations of brain or spinal injuries such as loss of dexterity and weakness and the positive phenomena such as increased tone and brisk reflexes as manifestations of a loss of descending motor control.

Central to any notion of motor control or spasticity is that of the 'final common path',

namely the lower motor neurone through which all reflex and voluntary commands are executed. Much past research has concentrated on elucidating the mechanisms by which the motor neurone pool in the spinal cord is recruited or activated under different conditions.

2.3.5.2 Tonic and phasic spinal mechanisms and the Henneman Size Principle.

On the basis of studies in the decerebrate cat, Granit and colleagues (1956) divided motor neurones into two groups, those producing large amplitude but brief discharges during stretch and those that responded with continuous small amplitude discharges during maintained stretch. The sustained discharges were augmented by a preceding tetanic stimulus and also by non-specific afferent inputs such as twisting the pinna of the ears or electrically stimulating the contralateral sural nerve: implying that these sustained discharges were state-dependent. In addition, deep anaesthesia abolished the tonic responses, but the phasic responses were much more resistant to anaesthesia. Accordingly, Granit *et al* (1956) postulated that two types of ventral horn cells were responsible for these observations. The possibility that these two systems were responsible for voluntary muscle activity was appealing: tonic cells effecting sustained postures and phasic cells effecting rapid, alternating and skilled movements. Lance, de Gail and Neilson (1966) used vibration to study this phenomenon, and found that a vibration-induced tonic contraction increased in force by recruiting motor units of increasing size as the vibration frequency was increased. During these studies, the maximum motor unit frequency, irrespective of size remained between 4 and 10/s The tonic contractions tended to be reduced or absent in lesions of the peripheral nerves or nerve roots, below the level of spinal lesions or brainstem lesions and in cerebellar and cerebral disease, including dystonic states and anoxic-decerebrate rigidity states. The tonic vibration response (TVR) was preserved in Parkinsonism. The TVR was also abolished by rapid stretching, but returned when this stretching ceased. The Authors postulated that synchronous afferent volleys would produce phasic discharges of large motor units and asynchronous discharges, would recruit smaller, lower threshold motor units.

This hierarchical recruitment from small through to increasingly large motor units was later elaborated into a now famous 'law of combination' (Hennemann *et al*, 1974) in which the motor system first (and always) recruits the smallest, motor units with low-firing thresholds which increase in rate of firing with increasing demands till further demands are met by recruiting larger motor units: producing the by now familiar 'diamond' EMG interference

pattern. As the demands for force generation lessens, so the motorneurone pool is de-recruited in the reverse manner, beginning with the largest, fastest and most fatigable motor units and ending with the smallest, slowest and least fatigable units.

These studies indicated a common pool of motor units, the elements of which are recruited on the basis of size: the smallest with the lowest thresholds first.

2.3.5.3 Relieving spasticity and releasing voluntary motor control.

Based on previous observations of the effects of the local anaesthetic agent, procaine, in abolishing brisk reflexes and clonus, Geoffrey Rushworth (1960), published a study of the effects of partial nerve blocks with local injections of dilute procaine anaesthetic aimed at selectively blocking small-diameter gamma fibres, the efferent motor nerves supplying the muscle spindles, in patients with a variety of spastic syndromes. Most of the 20 cases studied in this way appeared to gain strength after these infiltrations around the peripheral nerves. The explanation for the success of this treatment was based on the notion that phasic and tonic stretch reflexes competed with voluntary descending motor control for command of the lower motor neurone pool, but that if the reflex afferent input were diminished, by temporary chemical denervation of the efferent nerves to the muscle spindle (gamma fibres), greater voluntary control could ensue.

Injections in 25 cases of Parkinsonian rigidity, abolished the rigidity and preserved the voluntary power and likewise in 4 cases of dystonia , in which voluntary control was briefly enhanced (Rushworth, 1960). However, Landau, Weaver and Hornbein (1960) were unable to demonstrate improved coordination following differential nerve block studies.

Sahrman and Norton (1977), looked at the issue of phasic stretch reflexes impeding voluntary control at normal movement speeds. They found that healthy subjects could generate a reflex stretch response during rapid antagonist stretches, but that patients with an upper motor neurone syndrome could not move the limbs sufficiently rapidly to elicit a reflex response: in other words, the stretch reflex was not interfering with the little remaining voluntary control. McLellan (1977) also demonstrated reduction of passive stretch reflexes of the quadriceps and hamstrings after oral baclofen, but found that the co-contractions during voluntary sinusoidal tracking tasks were unaffected, indicating a persistence of the motor dyscontrol which appeared to be independent of the mechanisms supporting reflex excitability. In another study, McLellan, Hassan and Hodgson (1985) looked at position and

force tracking at the elbow at rates of 0.64Hz between 90° and 135° of elbow flexion in normal controls and spastic patients and found abnormal peaks of flexor EMG at peak elbow extension. These peaks also occurred in the flexor muscles during isometric extensor force tracking in which no elbow extension movements actually took place, so that the abnormal flexor EMG peaks could not be attributed to flexor muscle stretch reflexes, but instead , to abnormal motor planning. The administration of tizanidine abolished the passive stretch reflexes but did not affect the abnormal flexor muscle discharges at peak extension, in keeping with the earlier studies (McLellan, 1977).

2.3.5.4 Spasticity and velocity-dependent stretch reflexes.

Based on numerous observations in man (see Burke and Lance, 1973), Lance (1980) was able to formulate an operational definition of the abnormal stretch reflexes seen after brain and spinal injury in operational terms:

> " In both cerebral and spinal spasticity, the stretch reflex responses obtainable from extensor and flexor muscle groups of the upper and lower limbs increase approximately linearly with the increase in the velocity of stretch. The reflex component of the increased tone may therefore be measured in terms of the threshold velocity required to evoke reflex activity and the slope of the EMG-velocity relationship."
>
> *Lance, 1980.*

Similar velocity-dependent stretch reflexes were demonstrated by Tasker *et al* (1980) in the squirrel monkey, but this only became significant with bilateral ablations of the primary motor cortex, with no additional increases in spasticity after additional ablations to the supplementary motor cortex.

2.3.5.5 Spasticity and the pyramidal tracts.

The widespread notion that spasticity results from a pyramidal lesion has scant experimental support. 'Pyramidotomy' in monkeys produced a flaccid weakness with loss of distal fine motor control (Tower, 1940) but reflexes were not brisk. Nor was spasticity a feature in a reported case of pyramid destruction in an adult male (Bucy *et al*,1964). Also, as seen above, unilateral ablation of the precentral gyrus (area 4 of the cortex) also failed to produce spastic hypertonus (Tasker *et a*l, 1980). Indeed the commonest clinical experience is the development of severe spasticity in adults with acute capsular strokes which result in damage to pyramidal and extrapyramidal fibres (Denny-Brown, 1980; see also Brown, 1994, for a review)

2.3.5.6 Mechanisms of spasticity.

Considerable attention has been given to understanding the mechanisms of spastic hypertonus and the focus has moved away from that of dynamic muscle spindle hypersensitivity of the '50s and '60s, which was abandoned on the basis of microneurographic recordings from Ia afferents (see fig. 2.3 and Burke, 1983, for a review) which failed to demonstrate increased firing rates, in response to muscle stretches, in the spindle afferents. The implication followed that central abnormal processing of a normal stretch signal was a likely explanation for the lowered velocity thresholds to stretch and the increased velocity gain after cerebral or spinal injury and that presynaptic inhibition normally obtains in the healthy nervous system, but is lost or diminished to a varying degree after CNS damage (Burke and Ashby, 1972). This and other mechanisms have been reviewed by Katz and Rymer, (1989). Other central mechanisms have been advanced including sprouting of dorsal root collaterals (McCouch *et al*, 1958), although a recent study has failed to confirm the McCouch group findings (Nacimiento *et al*, 1993).

The principle site of action of Baclofen, a GABA-b receptor agonist is on the pre-synaptic nerve endings of the Ia afferent fibres in the dorsal horns of the spinal cord. (see section 1.3.5 .2 for detailed discussion and Bowery, 1989, Price *et al* , 1984, 1987).

2.3.5.7 Syndromic spasticity and the role of abnormal postures.

As was argued in section 1.3 3 on the classification of cerebral palsy, the elicitation of velocity-dependent reflexes plays only a small part of the syndromic definition as established by Crothers and Paine (1959) who included the release of postural and tonic labyrinthine reflexes, spread and overflow of associated movements, loss of voluntary control of fine finger movements and a tendency to muscular contractures: clearly these are the features of the whole motor disorder, not the spasticity which is a part of the motor disorder. The point is important because it returns to the original criticisms of medical practice raised by Landau (1974, 1988) who was concerned that hypertonus and more specifically, reflex hypertonus was receiving a disproportionate amount of attention to the detriment of issues of weakness and poor motor control (the negative phenomena).

Postures of one sort or another, as well as resisting passive stretch, form a necessary backdrop to all motor actions, whether voluntary or otherwise. They certainly seem to contribute considerably to the pathophysiology of equinus (section 3) and to regulate reflex

motor output (sections 5,6,7,8). They may play a role in voluntary motor output (section 9).

That they are conditioned by afferent input from the neck and the otoliths (see section 10 and Magnus and Kleijn, 1912; Walshe, 1923; Chan, 1983 for a review) and are strongly conditioned by voluntary (but unfamiliar) tasks (Føg and Føg, 1963), there is little doubt. Thus, position in space and a variety of nonspecific afferent stimuli will alter muscle tone via these postural mechanisms according to the state of maturity of the infant and child and in varying combinations with the severity of brain injury. What must be borne in mind is that mechanisms other than the muscle spindle afferent processing are responsible for disordered motor tone and function.

2.3.5.8 Spasticity, contracture and peripheral muscle transformation.

Herman (1970) has described contractures of the triceps surae muscle in his group IV hemiplegic adults who exhibited little reflex excitability and in whom clonus could not be elicited. Dietz, Quintern and Berger (1981) described alterations in the mechanical properties of muscle of children with cerebral palsy in whom the EMG recordings during the gait cycle were, if anything, diminished in amplitude (see Dietz, 1992 and Forrssberg and Dietz, 1997 for reviews). Hufschmidt and Mauritz (1985) also demonstrated an increase in the work of viscoelastic stretch of the calf muscles at velocities below the stretch reflex velocity threshold and consistent with normal walking speeds (ie <20°/s) keeping with peripheral muscular change: findings which were absent in Parkinsonism.

In contrast, O'Dwyer, Neilson and Nash (1994) attempted to influence the development of muscle contracture of the triceps surae by reducing the tonic stretch reflex gain in spastic children deemed to have a high level of spasticity. The programme involved three half-hourly sessions per week for 5 months and resulted in diminishing the tonic stretch reflex gain by some 50% or more using a feed-back technique but the muscle contracture was not altered when assessed by torque-angle curves. This suggests that factors other than the spasticity, *per se*, mediates muscle shortening.

2.3.6 Dystonia.

The issue of the presence or absence of dystonia in cerebral palsy is often debatable but was probably under-recognised in the past. Nevertheless, dystonia is increasingly recognised in the ex-preterm infant in whom it is more prevalent than the epidemiology of CP would lead one to believe (see Brown *et a*l, 1997). Dystonia is recognised as a state of

involuntary fluctuating postures, which by definition will offer , usually,marked resistance to passive stretch, and is entirely relieved by sleep (Fish *et al*, 1991 and see section 10 on postures). .

2.3.7 Tonic neck and tonic labyrinthine reflexes.

These will be defined in section 10, and their role in the understanding of moderate to severe diplegia will be reviewed. These posture-producing reflexes appear to have a much greater part to play in the motor difficulties of children with CP than is usually recognised.

2.3.8 Abnormal movement patterns.

These have been discussed in section 1, in the foregoing as part of the 'spastic syndrome' and are a feature of virtually all the subsequent sections, particularly sections 3, 6 and and 9 and 10. It is axiomatic that a disordered movement pattern results in inappropriate stiffness which might be mistaken for spasticity or other tone disorder, the problem being the pattern generator, not the reflex.

2.4. Summary.

An attempt has been made to review some of the basic mechanisms which contribute to abnormal muscle tone, inertial, viscoelastic and neurally-mediated, partly with a view to clarifying what we call things and how these entities may be observed and ultimately quantified., but upper most, to facilitate appropriate management.

As has been seen, the detail and complexity of motor organisation in health and disease is intricate. Nevertheless, some simple principles may emerge as guide-lines to managing a child with motor difficulties: the simplest of these being to consider in detail the various factors contributing to the total motor disorder.

3. Mechanisms of Hind-foot Equinus in Childhood Hemiplegia.

3.1 What is equinus?

Usually, walking patterns develop from a broad-based, flexed posture at the hip, knee and ankle around the age of one (with a wide normal age of onset) at which time there is marked co-contraction of the major lower-limb (Leonard, Hirschfeld and Forssberg, 1991), and presumably, trunk and upper-limb muscles. There follows from this largely triple-flexed, co-contracting pattern (figure 3.1, left), a transition to a more erect posture associated with a phasic interplay of the leg muscle agonists and antagonists which produce joint asymmetries during which, for instance, the hip and ankle joints may be flexed but the knee joint is extended, as for instance in terminal swing and at heel contact (figure 3.1.1, right).

This heel contact constitutes the hall-mark of a 'mature' gait pattern and is usually achieved around the age of 2 years (Sutherland, Ohlshen, Cooper and Woo, 1980; Sutherland, Ohlshen, Biden and Wyatt, 1988, p16), whereas the joint synchronies associated with co-contraction of muscles are a frequent accompaniment in children with cerebral palsy (Leonard, Hirschfeld and Forssberg, 1991).

In this context, the interplay between the gastrocnemius-soleus muscles posteriorly and the tibialis anterior muscles usually determines the foot-contact pattern according to the phases of the gait cycle in which they are active: ie 'stance phase' or 'swing phase'. In the mature gait, the gastrocnemius-soleus (G-S) muscle is only active after heel-strike, and ceases to be active at he beginning of plantarflexion prior to 'toe-off': this occurs in 33% of one-year old infants and 77% of two-year olds (Sutherland, 1966; Sutherland, Cooper and Daniel, 1980b; Sutherland, Ohlshen, Biden and Wyatt, 1988, p16 for phases of gait, p160-161 G-S muscle activity). An immature pattern is seen when the G-S muscles are active in 'terminal swing' and at foot-strike, which occurs in two thirds of infants aged 18 months and younger and less than 25% of toddlers of two years old over (Sutherland, Ohlshen, Biden and Wyatt, p160-161). The timing of the muscle activity is therefore a principal determinant of the pre-positioning of the foot prior to foot contact, and this timing of the muscle patterns is developmental, and may remain immature up to the age of seven in about a quarter of children (Sutherland, Ohlshen, Biden and Wyatt, 1988, p160-161). However, in these healthy children, the clinical examination is normal and the passive examination does not reveal excessive 'muscle tone' or brisk reflexes: what of children with motor disorders?

Figure 3.1 Developmental crouch stance (triple flexion) and mature heel-strike.
Left: Male infant aged 11 months with a developmental crouch posture: triple flexion at hip, knee and ankle joints, anterior pelvic tilt and hyperlordotic back.
Right: Same child, now a 23 month old toddler. Note: heel-strike foot-contact pattern, lower limb joint asymmetry (flexion of hip and ankle, but extension of knee joints respectively), lack of anterior pelvic tilt, straight back, reciprocating arm movements, gaze intent on destination.

A frequent clinical observation of children with hemiplegia and diplegia is that they walk on their toes: this is referred to as a 'toe-striking' or an 'equinus' gait pattern. In addition, these same limbs frequently offer increased resistance when the calf muscles are manually stretched by an examiner. It is therefore possible to say that such limbs exhibit both a 'gait equinus' when walking and a 'passive equinus' when clinically examined on the couch.

The clinical correlates of an abnormal foot-contact pattern are numerous and for each child it would be important to determine which are the primary determinants of the abnormal walking pattern and which are secondary phenomena. Table 3.1 indicates a variety of possible abnormalities which might, either singly or in combination, result in an equinus gait.

Table 3.1 Factors contributing to an equinus gait.

I. Electrically 'silent' mechanisms

- short leg
- short tendon: ie contracture
- short muscle: ie contracture
- stiff muscle: reduced compliance, altered viscoelasticity and electrically silent.

II. Electrically 'active' mechanisms

- foot-drop: i central due to weak dorsiflexors 'pyramidal weakness'
  ii peripheral
- phasic spasticity: a reflex equinus in terminal swing
- 'tonic spasticity': increased resistance to slow stretch
- dystonia: wide fluctuations in muscle tone
- equinus posture ie tonic labyrinthine release
- abnormal walking engram
- developmental immaturity
- elective or voluntary toe walking
- sprinting
- dancing

III. Combinations of I and II.

modified after Lin and Brown, 1992

As Elin and Mogens Føg (1963) have shown, developmental motor immaturity persists in up to a quarter of young adolescents up to the age of 16. Likewise, Sutherland and colleagues (1988) were surprised at the finding of motor immaturities in older children:

"Striking differences were found when the dynamic phasic activity of the gastroc-soleus in our pediatric population was contrasted with that in the normal adult control group (Inman *et al*, 1981). Swing-phase activity, not usually present in the adult, was commonly seen in our subjects, particularly in the younger age-groups. We found two patterns that require separate descriptions. The first we term the 'immature' pattern because it showed the greatest differences from that of the normal adult controls. It occurred in a very high percentage of the 1 and 1.5 year-olds and in approximately one-quarter of the subjects in the remaining age-groups. It is a wrap-around pattern, beginning near the middle of the swing phase and ending with the opposite foot-strike. 67 per cent of the 1-year-olds and 63 per cent of the 1.5 -year-olds demonstrated this immature pattern. There was a precipitous drop in the proportion of subjects with this pattern at 2 years; however, the finding that at 2 years and older, approximately one-quarter of our subjects still demonstrated this immature pattern was quite unexpected.

"In the second of these patterns (the "mature" pattern) activity of the gastroc-soleus is confined to the single-stance portion of stance phase, and therefore resembles the adult pattern (Sutherland, 1966; Sutherland *et al* ,1980b, Inman *et al* ,1981). The mature pattern was present in approximately one-third of the 1- and 1.5-year-olds and in around three-quarters of the older children"

Sutherland et al, 1988, p160

It is important not to mis-attribute to pathological patterns, what is in effect a developmental pattern of motor immaturity. For instance, the activation of the gastrocnemius-soleus muscles in terminal swing and foot strike could easily be misrepresented as a 'spastic' firing pattern producing an equinus foot-contact in a young child, whereas it may merely represent an immature motor pattern. Such developmental patterns of motor immaturity may be one reason why children thought to have 'moderate-to-severe' motor difficulties 'outgrew' their cerebral palsies (Nelson and Ellenberg, 1982). Similarly, such findings ought to guard the unwary against the use of highly invasive treatments such as 'selective dorsal rhizotomy' at increasingly younger ages, as has been advocated by some (Park and Owen, 1992).

Such muscle firing patterns need to be interpreted with caution since they may represent immaturity; may be compensatory for a 'physiological crouch' or some other motor pathology remote from the ankle joint, or, indeed, be the primary, pathological cause of the abnormal gait.

Clearly, the treatment (s) offered to a child exhibiting gait equinus, should vary according to the primary causes, which should be established wherever possible. For instance, it would be inappropriate to treat a gait equinus attributable to developmental immaturity or elective toe-walking with the same treatment as for a short tendon or muscle; or,

as if it were a 'spastic' out-of-phase muscle. This issue of a proper diagnostic attribution of the motor patterns is referred to below in sections 4 to 6.

3.2 Methods

3.2.1 Patients

A convenience sample of 24 children with hemiparesis or hemiplegia attending the Motor Clinic at the Royal Hospital for Sick Children, Edinburgh were consecutively invited to take part in a non-invasive clinical study.

Table 3.2.1 gives the salient clinical details of these 24 children including age, sex, attributed cause according to whether the hemisyndrome was considered to be congenital, perinatal or acquired; side of hemisyndrome; presence or absence of seizures; requirement for special education; gait grade and finally according to the patterns of 'muscle tone' exhibited: ie phasic spasticity, tonic spasticity, dystonia or normal tone. The pattern of walking according to the foot-contact pattern based on a score from 1-4 as follows: 1: heel strike; 2: plantar strike; 3: toe-heel; 4: toe-strike (equinus) gaits respectively. The ages spanned 6-17 years (mean 9.9 years). Only four of the children had hemisyndromes attributable to post perinatal causes, reflecting the balance of distribution of causes within the clinic population as a whole.

3.2.2 Measurements of clinically assessed variables.

These included **i.** leg length (cm), **ii.** ankle joint range (°), **iii.** extensibilité (°), **iv.** compliance difference (°/unit torque), **v.** gait score, **vi**. Medical Research Council (MRC) muscle power grade for the dorsiflexor and plantarflexor muscles (grade 0-5),**vii** distal dexterity of the toes (Hz) and **viii** clinical patterns of muscle tone: these variables are summarised in table 3.2.2. It should be noted that this study represented no more than a carefully documented clinical examination in which the clinical measurements were operationally defined (see section 2 above).

Table 3.2.2 Clinically assessed Variables

	Variable	Units
i.	Leg length difference	cm
ii.	Ankle joint range	degrees (°)
iii.	Extensibilité*	degrees (°)
iv.	Compliance difference	degrees/unit torque (absolute torque varying from subject to subject)
v.	Gait score:	grade 1 = heel strike grade 2 = plantar strike grade 3 = toe-heel strike and back-knee grade 4 = obligate toe strike
vi.	MRC# power scale	grade 0 = no contraction grade 1 = flicker of contraction grade 2 = movement, gravity eliminated grade 3 = movement against gravity grade 4 = movement against resistance grade 5 = Normal power
vii.	Distal dexterity of toes	frequency of alternating flexion/extension in Hz
viii.	Clinical muscle tone	phasic spasticity: resistance to rapid stretch tonic spasticity: resistance to slow stretch dystonia: widely fluctuating co-contraction rigidity: uniform, unvarying co-contraction

* Extensibilité: that portion of the total joint range attributable to muscle lengthening.
\# MRC: Medical Research Council.(1976) *Aids to the examination of the peripheral nervous system.* Memorandum N° 45. London: Her Majesty's Stationary Office.

i. Leg length.

The leg length difference between the normal and affected limb was recorded in centimetres.

ii. Ankle joint range.

Ankle joint ranges in degrees were obtained for normal and affected limbs and the difference recorded. Joint angles were assessed by visual inspection of the sole of the foot in relation to the shaft of the tibia. By convention, neutral was defined as the position when the sole is perpendicular to the tibia and the angles of dorsiflexion and plantarflexion measured relative to the neutral position. The position of resting equinus was measured for normal and affected sides in all cases with the subject supine.

iii. Muscle extensibilité.

Extensibilité difference was measured in degrees. The term "extensibilité" was coined by André-Thomas (1949) to define that portion of the total joint range corresponding to muscle lengthening. Joint angles were assessed visually. The tension corresponding to muscle lengthening was gauged by the examiner. The angle from the first onset of tension in the tendon till the onset of a further steep rise in tension was measured and represented the extensibilité of the muscle in degrees.

iv. Compliance difference.

This was measured with the child supine. Equal pressure was simultaneously applied to the soles of the feet (a 'unit torque'): the difference in angular displacement at the ankle joint then represents the compliance difference in degrees/unit torque (fig. 3.2.1).

v. Gait score.

Gait was scored on a 1-4 scale which is detailed in table 3.2.2:

1: heel strike; 2: plantar strike; 3: toe-heel; 4: toe-strike (equinus) gaits respectively.

vi. Power grade.

The Medical Research Council (MRC memorandum N° 45, 1976) muscle power grades were used. Muscles were tested isometrically. A muscle was said to have normal power if no difference could be found between affected and unaffected limbs. Power was tested with the subject supine and in the case of the plantarflexor and dorsiflexor muscle groups, with the knee fully extended. In each case, the optimal maximum voluntary contraction sensed by the examiner was used as the final score with no attempt at standardising the the ankle joint position since the children positioned their ankle joints at a self-selected and presumably, most favourable angle.

vii. Motor dexterity.

Fine motor dexterity was obtained by measuring the maximum frequency of alternating flexion and extension of the toes in Herz (Hz).

viii. Definitions and assessment of muscle tone.

Muscle tone was defined as the resistance felt as a limb is passively rotated about a joint at rest (see section 2, above). *Phasic spasticity* was used to describe the resistance to rapid stretch. *Tonic spasticity* the resistance to slow stretch. *Dystonia* was used to describe wide fluctuations in tone due to co-contraction of muscles with changes of body positions in

space, and in response to non-specific afferent stimuli such as conversation, noise and mental arithmetic etc., together with bizarre, sustained postures. Muscle tone was assessed clinically according to whether it was *normal*, or showed evidence of *phasic*, *tonic* or mixed *phasic/tonic* spasticity or of hemidystonia.

3.2.3 Definitions and measurement of hind-foot equinus.

Two assumptions were made:

a The examiner can judge a joint angle to within 5°and **b.** the examiner can balance the force applied to dorsiflex the ankles of both legs simultaneously and apply similar forces to both sides. This is based on Foley's (1960) observations of the examiner being able to accurately sense differences in muscle tension transmitted to the examiner's limbs. Based on these assumptions, hind-foot equinus was defined in two ways:

1. Gait equinus: an obligate toe strike maintained throughout stance in the gait cycle.
2. Passive equinus: a compliance difference of greater than 10°/unit torque in passive dorsiflexion.

3.2.4 Statistics.

Differences between means were assessed using the unpaired Student's t test. The chi-square test was used to determine differences between groups and the unpaired Wilcoxon Rank Sum test, W, for the analysis of differences in ordinal grades of power between groups.

3.3 Results

Figure 3.3.1 represents 3 children with different forms of congenital hemiplegia The top row of pictures show a six year old boy (case 17) with a right hemiparesis walking; supine resting plantarflexion (resting equinus); attempting active voluntary dorsiflexion and finally being passively dorsiflexed by the examiner. The resting equinus appears to be similar at both ankles, but active dorsiflexion is diminished on the right as is passive dorsiflexion when the examiner attempts to apply equivalent dorsiflexing torques to both sides.

The middle row illustrates the same sequence for a 9 year old girl (case 6) with an upper-limb dominant right hemiparesis: in this case, she exhibits a mature heel-strike pattern of foot-contact. In this case too, the resting equinus is similar on both sides but active and passive dorsiflexion are reduced on the right compared with the left. The right foot also appears to be mildly supinated ie inverted.

The last row of figure 3.1, depicts the case of a 12 year old girl (case 23) who exhibits hemidystonia (see also fig. 1.3.3 above): the left wrist is extended and the hand fisted, while the left foot makes heel-contact with the ground. Resting equinus is similar at both ankles, though note the spontaneously extended left toe, which is a hall-mark of dystonia. Active and passive dorsiflexion are reduced on the left compared with the right. In all three children, the signs were strictly unilateral and the back was straight. Only one of these children exhibited gait equinus despite the fact that all three appeared to have evidence of passive equinus on passive dorsiflexion.

Table 3.3.1 summarises the main findings for all 24 cases of hemiplegia according to equinus and non-equinus categories. There were no cases of fixed equinus among the 24 hemiplegic children: all affected ankle joints could be passively dorsiflexed beyond neutral. Overall, the ankle joint range was statistically significantly reduced on the hemiplegic side compared with the normal limb ($p < 0.03$). Normal and affected limbs had similar angles of resting equinus with the child supine. The resting angle of the affected limb is not helpful in predicting which hemiplegic limb will have an equinus gait.

The resting equinus for normal and affected limbs in left hemiplegia was significantly less than the resting equinus on the normal and affected sides for right hemiplegia, a finding for which there is no explanation. Details of all the goniometry may be found in Tables 3.3.2. and 3.2.3.

Legend to figure 3.3.1
Gait equinus, resting equinus and passive equinus in congenital hemiplegia.

see landscape figure overleaf (p93)

(top) Case 17: 6 year old boy with congenital right hemiplegia and gait equinus (toe strike).
(centre) Case 6: 9 year old girl with congenital right hemiplegia and a heel strike.
(lower) Case 23: 12 year old girl with a congenital left hemidystonia secondary to a discreet right pallidal infarct showing a normal heel strike.

i. Walking: foot-contact patterns
ii. Supine resting equinus in each case is similar for normal and affected sides, irrespective of power ranges or muscle tone.
iii. Active dorsiflexion brings out the weakness of the affected limb for case 17.
iv All had passive equinus, a compliance difference of greater than 10°/unit torque: equal pressure is applied to the soles of the feet and the difference in angular displacement is the compliance difference in degrees/unit torque.
v. For the group as a whole, gait equinus could occur in the absence of weakness and heel-strike could occur in the presence of short, clinically spastic legs.

After Lin and Brown 1992, by permission.

Figure 3.3.1

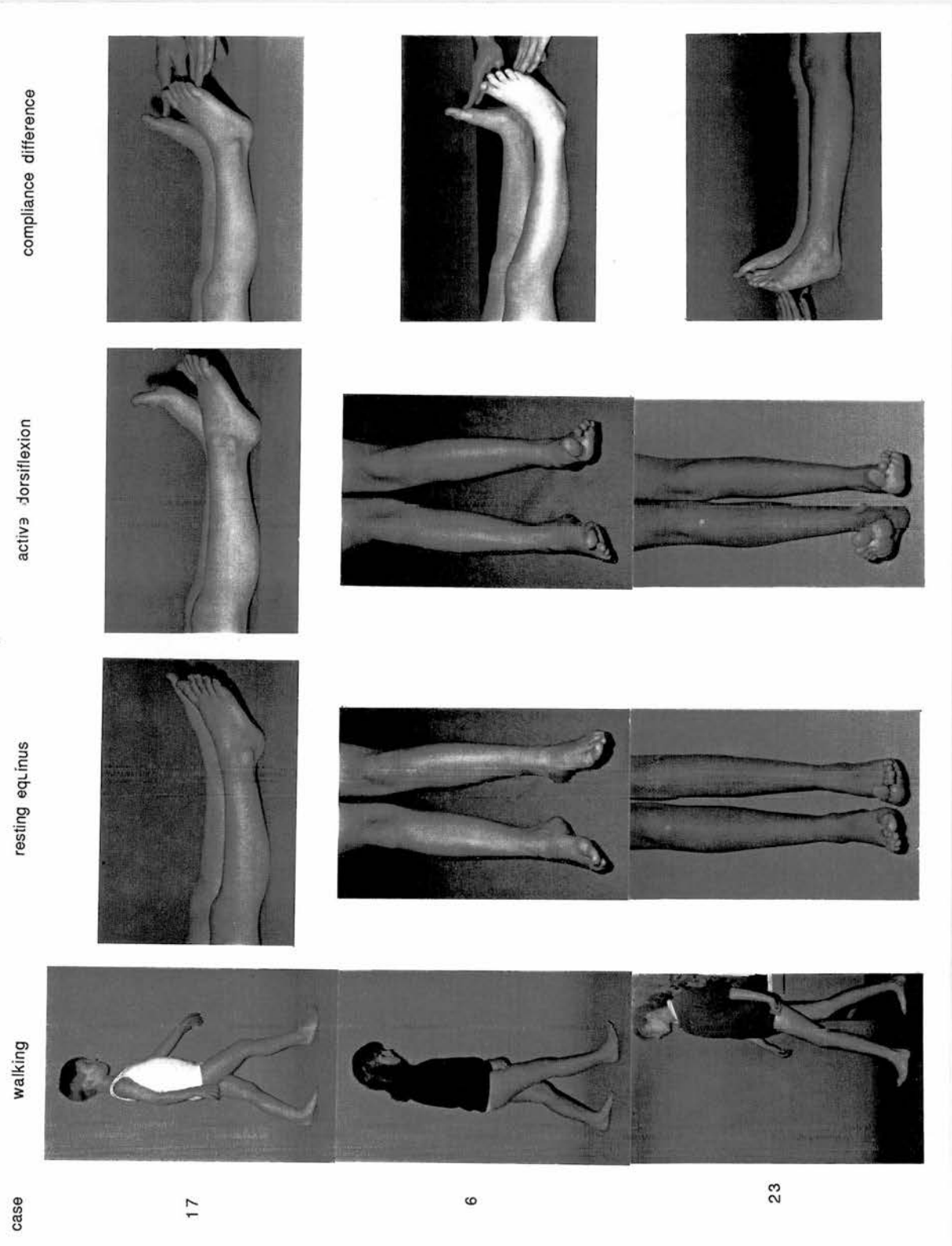

compliance difference
active dorsiflexion
resting equinus
walking
case
17
6
23

Table 3.3.1 Differences between equinus and non-equinus groups as defined by gait and compliance difference in childhood hemiplegia .n = 24.

Variable	Gait equinus Yes n=9 mean(SD)	Gait equinus No n=15 mean(SD)	Statistics.	Passive equinus Yes n=16 mean(SD)	Passive equinus No n=8 mean(SD)	Statistics
Leg length difference (cm)	2.0 (1.1)	1.8 (1.4)	NS*	2.2 (1.2)	1.1 (1.0)	P<0.03*
Ankle joint range difference (°)	12.5 (8.0)	8.0 (7.7)	NS*	12.5 (7.3)	2.9 (4.9)	p<0.006*
Extensibilité difference (°)	10.0 (6.1)	8.7)	NS*	12.2 (8.2)	3.1 (5.3)	p<0.02*
Compliance difference (°/unit torque)	23.9 (9.6)	14.5 (13.1)	NS*	----------		-
Toe dexterity (Hz)	2.1 (0.5)	2.4 (0.8)	NS*	2.3 (0.8)	2.2 (0.5)	NS*
Hemiside						
R	7	7		8	6	
L	2	8	NS**	8	2	NS**
MRC Power						
plantarflexion	----------		NS***	----------		NS***
Dorsiflexion	----------		NS***	----------		NS***
Clinical tone						
Phasic No	3	8		8	6	
spast.. Yes	6	7	NS***	8	2	NS***
Tonic No	4	6		4	6	
spast.. Yes	5	9	NS***	12	2	p<0.02***
dystonia						
No	8	14		14	8	
Yes	1	1	NS***	2	0	NS***

All mean differences are calculated between the normal and affected limb.
MRC = Medical Research Council Memorandum N° 45, 1976. *Aids to the examination of the peripheral nervous system.* Note no entries for power grades since these are ordinal numbers which cannot be handled parametrically.
No compliance differences given for passive equinus since it defines passive equinus.
NS = Not Significant. * = unpaired Student's t test; ** = Chi-square test; ***= unpaired Wilcoxon rank sum test.

Modified after Lin and Brown, 1992.

Table 3.3.2 Goniometry (°): Nonparetic versus Hemiparetic Side.

Variable	Nonparetic mean(SD)	Hemiparetic mean(SD)	t test
Ankle joint range	57.6 (6.1)	53.7(6.1)	<0.03

Modified after Lin and Brown, 1992, by permission.

Table 3.3.3 Goniometry (°): Right and Left Hemiplegia

	Rt hemiankle	Lt nonparetic	t test	Rt nonparetic	Lt hemiankle	t test
Joint	52.5 (6.1)	58.9 (4.9)	p<0.006	63.3 (9.7)	55.6 (5.8)	NS
range	52.5 (6.1)				55.6 (5.8)	NS
		58.9 (4.9)		63.3 (9.7)		NS
Resting	35.7 (11.1)	32.1 (7.30	NS	14.6 (18.6)	16.2 (20.6)	NS
equinus	35.7 (11.1)				16.2 (20.6)	P<0.003
		32.1 (7.3)		14.6 (18.6)		P<0.003

Modified after Lin and Brown, 1992, by permission.

Gait equinus and non-equinus gait.

There were 9/24 cases of gait equinus. Of the remaining 15/24 cases: 3/24 had heel-strike, 7/24 a plantar-strike and 5/24 a toe-heel gait (fig 3.3.1, above).

A comparison of the clinically assessed variables between the gait equinus and non-equinus groups showed no statistical differences in leg length, ankle joint range, extensibilité, fine motor dexterity, the side of hemiplegia, MRC power grading of plantarflexors and dorsiflexors or of muscle imbalance across the ankle joint, nor any differences in the muscle tone at rest of the affected limbs in either group (table 3.3.1, above). Although 8/9 cases of gait equinus had compliance differences which satisfied the criteria for passive equinus there was no overall difference in compliance difference between the gait equinus group and those with other gaits.

Passive equinus versus no passive equinus limbs.

By contrast, the 16/24 cases with passive equinus (a compliance difference of more than 10°/unit torque) had statistically shorter legs ($p<0.03$), reduced ankle joint ranges ($p<0.006$), and reduced extensibilité ($p<0.02$) together with a clinical impression of 'tonic spasticity' ($p<0.02$) of the affected limbs compared with the affected limbs of those with normal compliance. Overall, 12/14 of all those with clinical evidence of 'tonic spasticity' had criteria for passive equinus. There were no differences in fine motor dexterity, the side of hemiplegia, or in the MRC power grading of the affected limbs in either group (table 3.3.1).

MRC power grades.

Similar trends of muscle imbalance are apparent irrespective of group category. The isometric power at the ankle has been plotted in order of increasing imbalance in figure 3.3.2**a-d** for gait equinus 3.3.**2a**, passive equinus 3.3.**2c** and their respective non-equinus groups. The actual plantarflexor and dorsiflexor power values have been superimposed on the imbalance rankings in figures 3.3.**2b** for gait equinus and 3.3.**2d** for passive equinus.

There was an unexpected finding of apparently normal isometric power in 6/24 cases. Of these six children, cases 4 and 21 had both gait and passive equinus, cases 22 and 23 had passive equinus alone and cases 6 and 10 had no equinus by either definition. The lack of weakness in cases 21 and 23 can be explained by both cases having a hemidystonic syndrome in which there was no muscle wasting, power of the anterior and posterior compartments was preserved, but functional impairment resulted from strong involuntary co-contractions, presumably resulting in a "subtraction weakness" due to opposing forces cancelling each other out. Another singular finding was that of *stronger power* in the anterior rather than posterior compartment in case 7, which accounts for an imbalance score of "-1" in figure 3.3.2.

Figure 3.3.2a-b MRC power grade s and the presence of gait equinus.

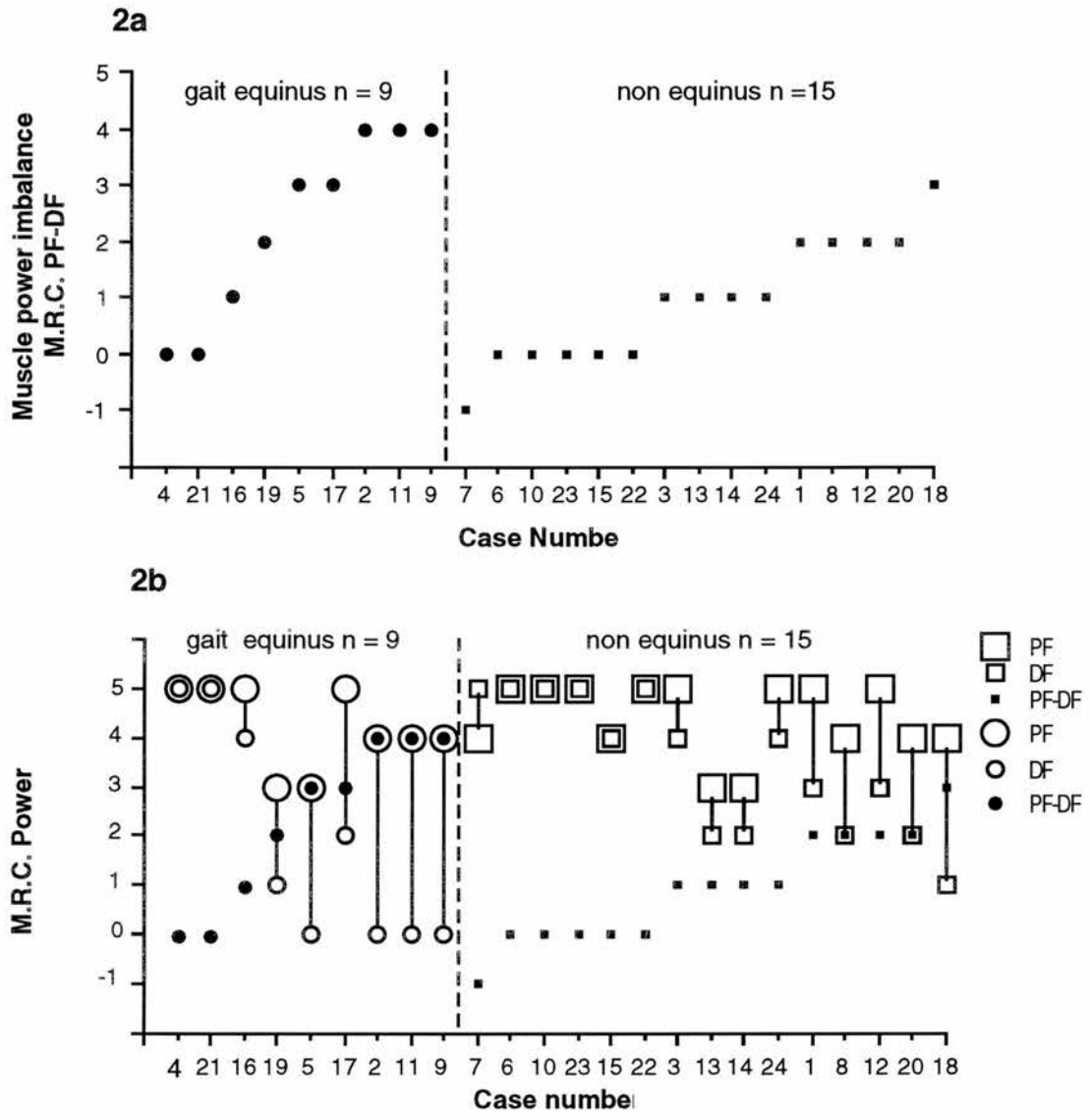
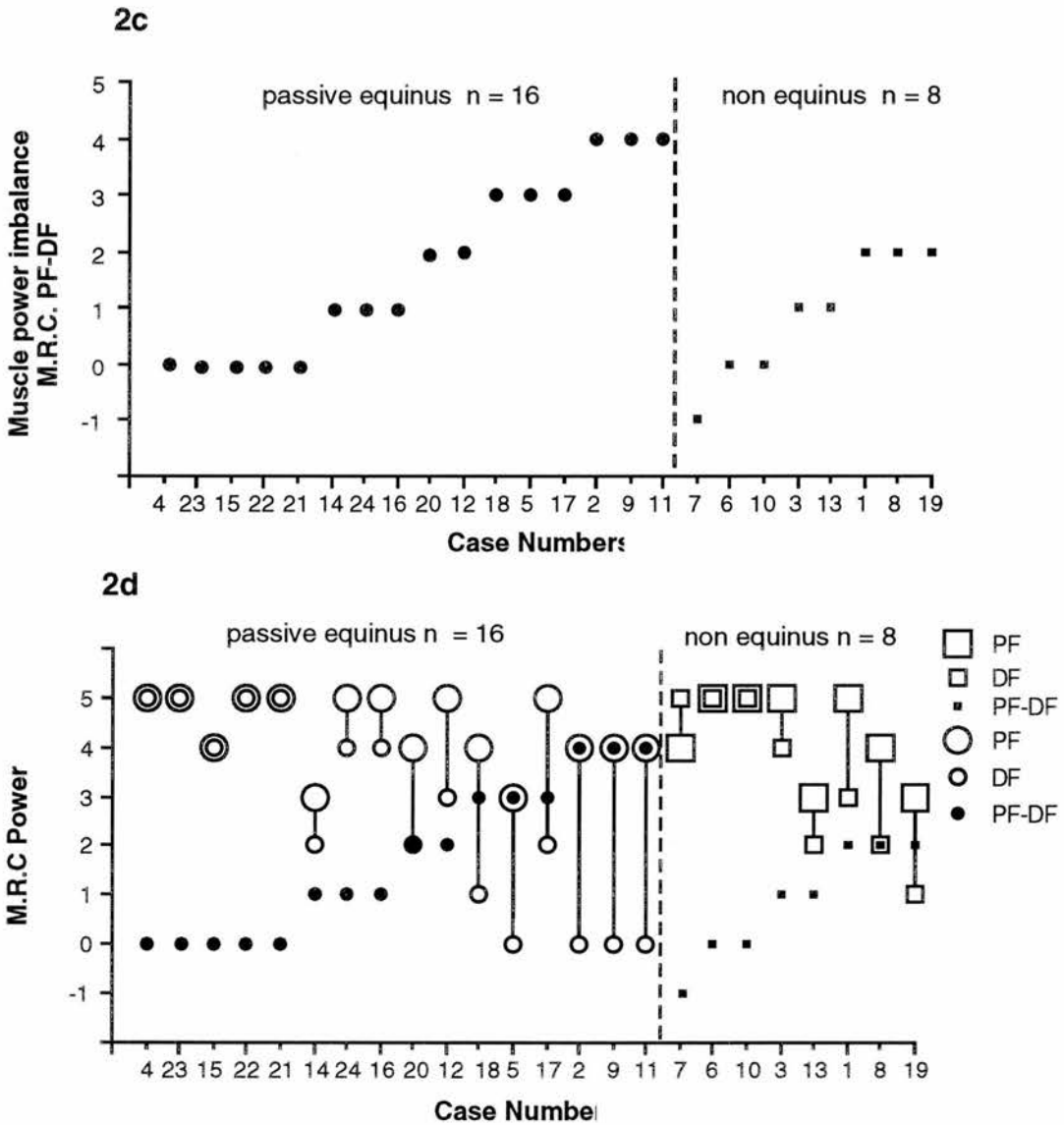


Figure 3.3.2c-d MRC power grade s and the presence of passive equinus.



The 'imbalance score'=PF-DF (solid symbols) indicate the ranking of imbalance from: 0=none to 4=marked imbalance. Note case 7: plantarflexors are actually weaker than dorsiflexors.

3.4 Discussion and interpretation of findings.

3.4.1 Heterogeneity of the hemiplegic population.

Subtle differences in the clinical expression of brain damage can be gauged with clinical experience by plain observation of the hemiplegic child. Instrumentation is not required and may obscure important pattern differences. However, once these clinical differences have been elicited, more detailed study is necessary to understand their possible mechanisms.

In previous studies the hemiplegic children have been considered as a homogeneous group (see Brown *et al* 1987, Brown *et al* 1991), the normal and affected limbs being compared using quantitative measurements of limb function. In contrast to these, the present study recognises the hemiplegias as heterogeneous and attempts to unravel those factors which result in different signs and motor developments of the affected side by distinguishing those children with and without a functional impairment i.e. gait equinus. This study sought to determine whether an operational definition of hind-foot equinus such as passive equinus, elicited during the clinical examination, correlated with a functional impairment such as gait equinus and other peripheral variables.

3.4.2 Goniometry.

All hemiplegic limbs had a significantly reduced total passive ankle joint range ($p < 0.03$), though differences were small and did not constitute a fixed deformity. The right hemiplegias had the largest differences in ankle joint range ($p < 0.006$), table 3.3.2, but none of these joint range differences could account for differences in gait. The resting equinus angles of normal and affected limbs were similar, giving no clue to which cases had either a gait or a passive equinus. An unexplained finding was that the resting equinus of normal and affected limbs in left hemiplegia was half that found in right hemiplegias (table 3.3.2). Resting equinus cannot explain the differences in gait patterns found in the 24 children.

3.4.3 Gait equinus, leg length, ankle joint range and extensibilité.

There were 9/24 cases with an obligate toe strike and toe stance. This contrasts with a study using foot-switch contact patterns in which no equinus gaits were found in the hemiplegic group (Csongradi *et al* 1979) though only two hemiplegic limbs had been observed. A heel strike occurred in only 3/24 of the presented cases, testifying to the overall abnormality of gait on the affected side in this hemiplegic group..

It had been anticipated that gait equinus would be associated with a compensatory equinus secondary to limb hemiatrophy but such a mechanism could not be confirmed despite a mean leg length difference between normal and affected limbs of 1.9 cm (SD 1.3 cm, range 0-4cm). Subjects with a unilateral short leg but no neurological signs walk with a Trendelenberg (waddling) gait, not a compensatory hindfoot equinus. There was no association with other peripheral factors such as reduced ankle joint ranges or limited muscle extensibilité, which supports the thesis that these peripheral changes do not necessarily cause gait equinus, but are the consequences of a common lesion.

3.4.4 Gait equinus, muscle power and the "classical" power model of equinus.

The lack of a clear correlation with dorsiflexor weakness, equivalent to a central foot drop, or muscle imbalance due to reciprocal inhibition at the ankle joint, was the most surprising result for the gait equinus group. Gait equinus is more complex than a simple central paralytic foot drop (table 3.3.1, figs. 3.3.2a-b).

These results appear to conflict with those of Sharrard (1964) who looked at the distribution of equinus deformity in relation to a relative paresis of dorsiflexion in 104 spastic limbs. The major differences between Sharrard's data and the present data being that his group was not restricted to hemiplegia. Sharrard's results included adult cases and he did not define what he meant by "equinus deformity". In Sharrard's data, a severe equinus could occur in the presence of mild or absent muscle imbalance.

The classical teaching on the mechanisms of deformity embraced four basic premises to explain evolving muscle imbalance at the ankle joint (fig. 3.4.1) which represented a synthesis of the known mechanisms of deformity following lower motor neurone lesions, ie polio and spina bifida, and the inferred pathophysiology of spastic states. This view of equinus is dominated by the concepts of muscle weakness and imbalance (fig 3.4.1) as opposed to abnormal muscle activation or temporal sequencing patterns (fig.3.4.2).

The first premise is that central weakness, arising from reduced corticospinal input with relative imbalance of the dorsiflexor to calf muscles, is a cause of gait equinus. Bleck (1987) has stated that there is normally a difference in torque between the gastrocnemius-soleus complex and the anterior compartment muscles acting to dorsiflex the foot at the ankle, leading many to wonder why a normal individual does not develop an equinus on the basis of a normal physiological muscle imbalance. All the children presented had similar

degrees of *resting* equinus on the normal and hemiplegic sides: that is to say, equinus in the absence of any conspicuous muscle activation.

The second premise relates to 'spasticity' of the calf, which has been considered as a cause of 'tonic reciprocal inhibition' of dorsiflexor agonist contraction, exacerbating the central dorsiflexor weakness by a peripheral spinal mechanism leading to further muscle imbalance across the ankle joint. This peripheral dorsiflexor weakness was thought to be proportional to the severity of the 'spasticity'. Sharrard taught that gait improved if the muscle imbalance was restored and this could be achieved by weakening the calf and removing the 'spasticity' through unloading the stretch on the calf. In other words, according to this theory, the constant stretch on the calf muscles produced a tonic contraction which was responsible for the equinus. (No evidence of tonic muscle activity was seen under EMG in section 6, below).

The third premise, an extension of the second involving 'spasticity', related to the observation that power in the dorsiflexors varies according to the position of the knee joint, the angle of which determines the tension, and hence spasticity in the gastrocnemius muscle. The power of the dorsiflexors can thus be unmasked if the knee joint is flexed, the calf muscles unloaded and the 'spasticity' relieved (Silverskjöld, 1923). This has led to some differences in opinion among orthopaedic surgeons about whether power could be accurately tested in upper motor neuron syndromes in the presence of 'spasticity'.

The fourth premise was failure of muscle growth. Deformities arise much more commonly during the growing period. Normally the muscles on each side of a joint grow to accommodate bone growth. If the epiphysis is stapled, the muscles readjust to the new limb growth potential (Sharrard, 1974). A muscle will grow longer and add sarcomeres if stretched, as in the case of the tibialis anterior, or shorten, removing sarcomeres in the gastrocnemius-soleus if no opposition or forced-lengthening occurs (Tardieu *et al* ,1988). If the dorsiflexors are weak, there is no stretch to prevent the soleus-gastrocnemius complex staying at its shortest length. This results in failure of normal growth, a reduction in the number of sarcomeres with a true shortening of the muscle: ie a fixed muscular contracture.

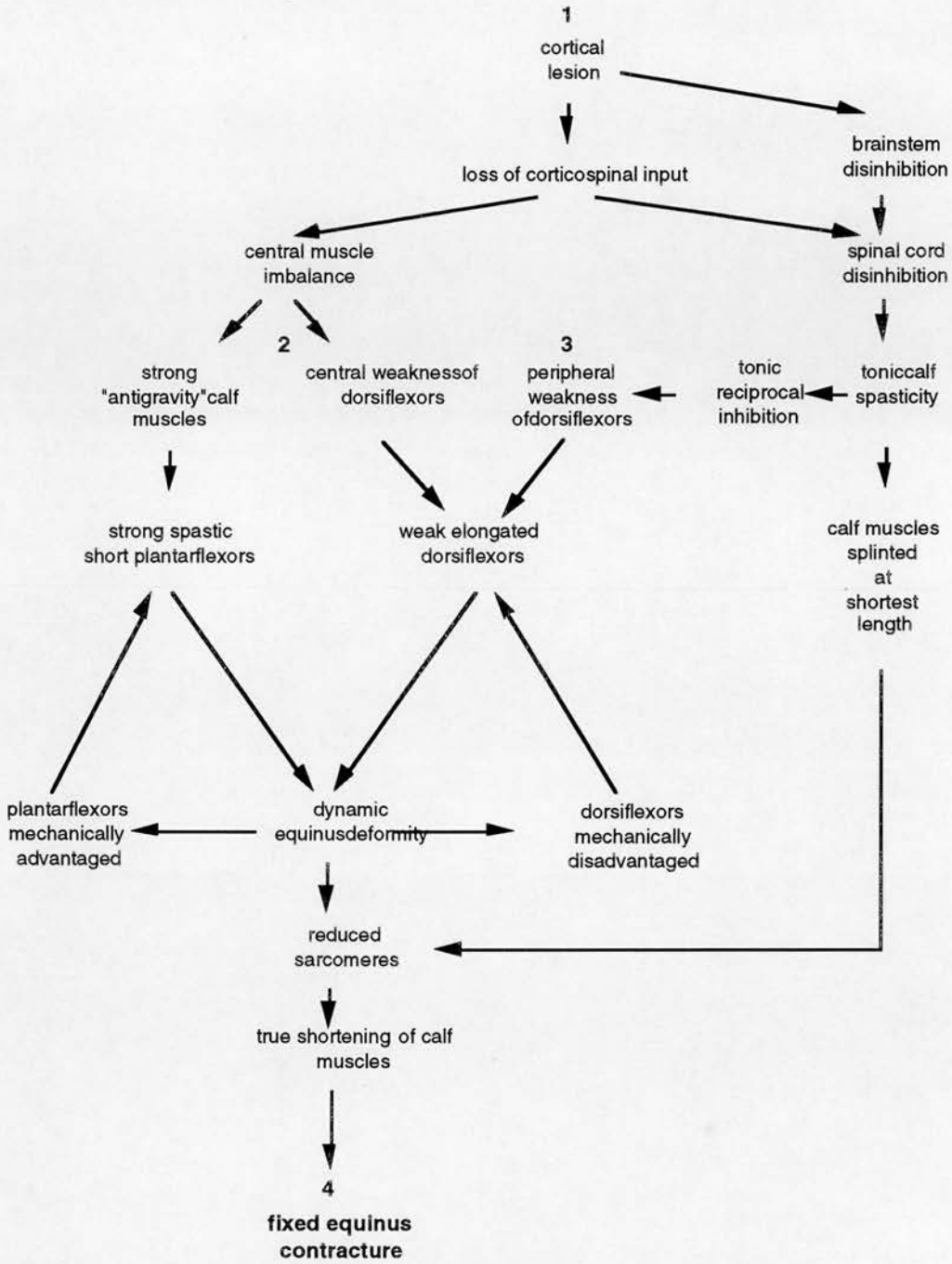


Figure 3.4.1.Classical Muscle Power Model of Equinus Deformity.
1. A cortical lesion results in central weakness and spasticity. 2. A central imbalance at the ankle produces strong,"antigravity" calf muscles and weak dorsiflexors. 3. Calf spasticity causes tonic reciprocal inhibition of dorsiflexors and peripheral weakness. 1, 2 and 3 combine to produce strong, spastic, short plantarflexors and weak elongated dorsiflexors resulting in an equinus posture. The equinus posture was thought to place calf muscles at a mechanical advantage based on the belief that the optimum power occurs at the resting length of the muscle. In equinus, the dorsiflexors were thought to be at a gross mechanicaldisadvantage. A vicious cycle sustains the equinus posture resulting in eventual true shortening of the calf muscles and a fixed equinus contracture (4). This scheme rests heavily on the notion of weak dorsiflexors and "strong" plantarflexors.

Central paresis with muscle imbalance, 'spasticity' and reciprocal inhibition seemed logical and interdependent mechanisms for the production of a short muscle and a fixed contracture (fig. 3.4.1). This classical scheme can be modified to take into account the non-neurogenic phenomenon of increased peripheral muscular stiffness demonstrated by Foley (1961) to be independent of reflex activity.

In practice, the clinical assessment of power is confounded by the fact that there is an optimal ankle joint position for dorsiflexor and plantarflexor maximum voluntary contractions (MVC). For dorsiflexion in normal adult subjects, this MVC angle corresponds to -10° of plantar flexion from neutral decreasing sharply as dorsiflexion increases beyond -5° of plantarflexion (Marsh *et al*,1981) whereas plantarflexor MVC occurs at +15° to +20° of dorsiflexion, being reduced five-fold at -30° plantarflexion. The position of the knee alters the angle of plantarflexor MVC, shifting it towards +5° of dorsiflexion with full knee extension or to +20° of dorsiflexion with 90° of knee flexion (Sale *et al* 1982). There is clearly a physiological range of ankle joint positions at which anterior and posterior muscular torques at the ankle joint are optimal (fig. 3.4.3a-b) and these do not correspond to the resting lengths of the dorsiflexors or plantarflexors. The tibialis anterior is stronger at -30° of plantarflexion than in the neutral position and plantarflexors are weaker with increasing plantarflexion. This finding suggests that the equinus posture favours the dorsiflexors to the detriment of plantarflexion and that plantarflexors are weakest in equinus: a complete refutation of classical teaching.

These length-tension characteristics clearly have implications for assessment of muscle power in normal and cerebral-palsied children as well as therapeutic implications. The MRC memorandum N° 45 "Aids to the examination of the peripheral nervous system" shows gastrocnemius being tested in a position of virtual maximum plantarflexion, ie a position of maximum weakness, while tibialis anterior is assessed at the neutral position: positions now known to be sub-optimal for MVC. For most of the children in this study, the ankle joint was allowed to alter to the position of optimum isometric MVC for that child when testing both plantar and dorsiflexor power.

Since actual walking does not involve isometric contraction, extrapolation from static length-tension relations must be treated with caution. Nevertheless the newer evidence on ankle mechanics upsets older concepts regarding the pathophysiology of equinus but accommodates the notion of abnormal central motor activation patterns.

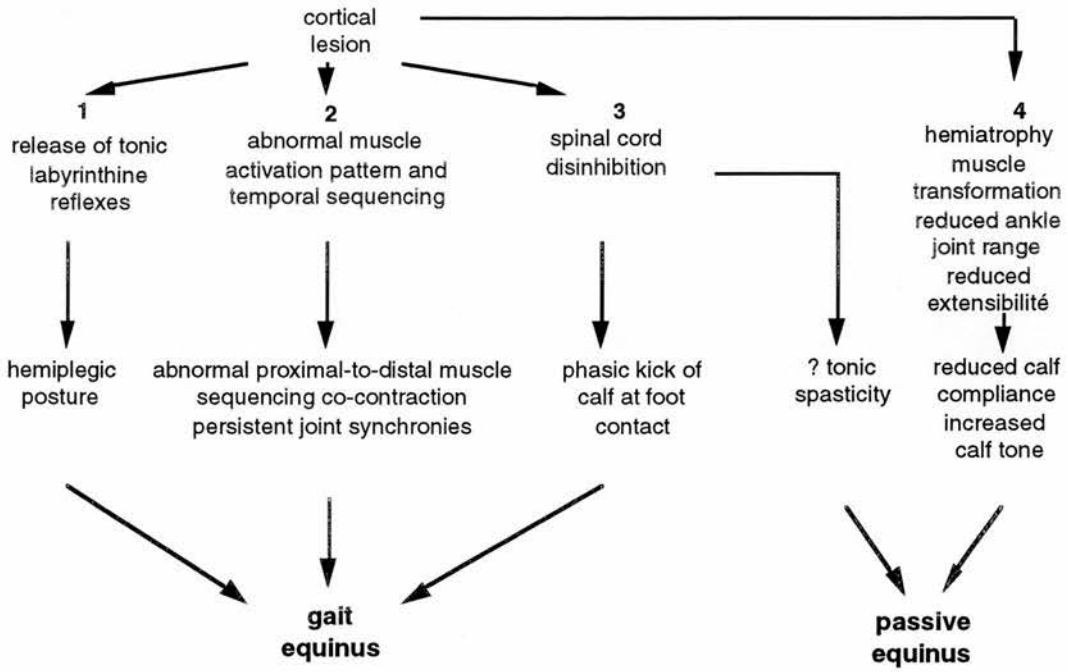


Figure 3.4.2 Developmental Model of Equinus in Hemiplegia.
1. A cortical lesion releases tonic labyrythine reflexes producing a hemiplegic posture. 2. Abnormal central muscular temporal sequencing patterns develop. This causes an abnormal proximal-to-distal muscle activation in the lower limb, persistent co-contraction, persistent joint synchronies and a failure of acquisition of the mature "heel strike". 3. Spinal disinhibition results in a phasic "kick" of plantarflexors at "toe strike" and possible tonic spasticity of calf muscles. 1, 2 and 3 combine to produce a dynamic equinus gait which is independent of absolute power ranges, leg length or muscle tone. 4. The cortical lesion results in variable degrees of hemiatrophy and muscular bio-transformation. The bio-transformation is associated with reduced extensibilité, reduced ankle joint range and increased resistance to passive stretch at rest producing a passive equinus.

After Lin and Brown, 1992, by permission.

3.4.5 Gait equinus and muscle tone.

Gait equinus could not be explained in terms of increases in muscle tone in the form of tonic spasticity as conventionally defined when clinically assessed at rest. All but one of the gait equinus cases had passive equinus but only half of the cases of passive equinus had an equinus gait, indicating that passive equinus is a necessary but not a sufficient cause of gait equinus. Factors other than a simple reduction in calf muscle compliance is required to produce a gait equinus.

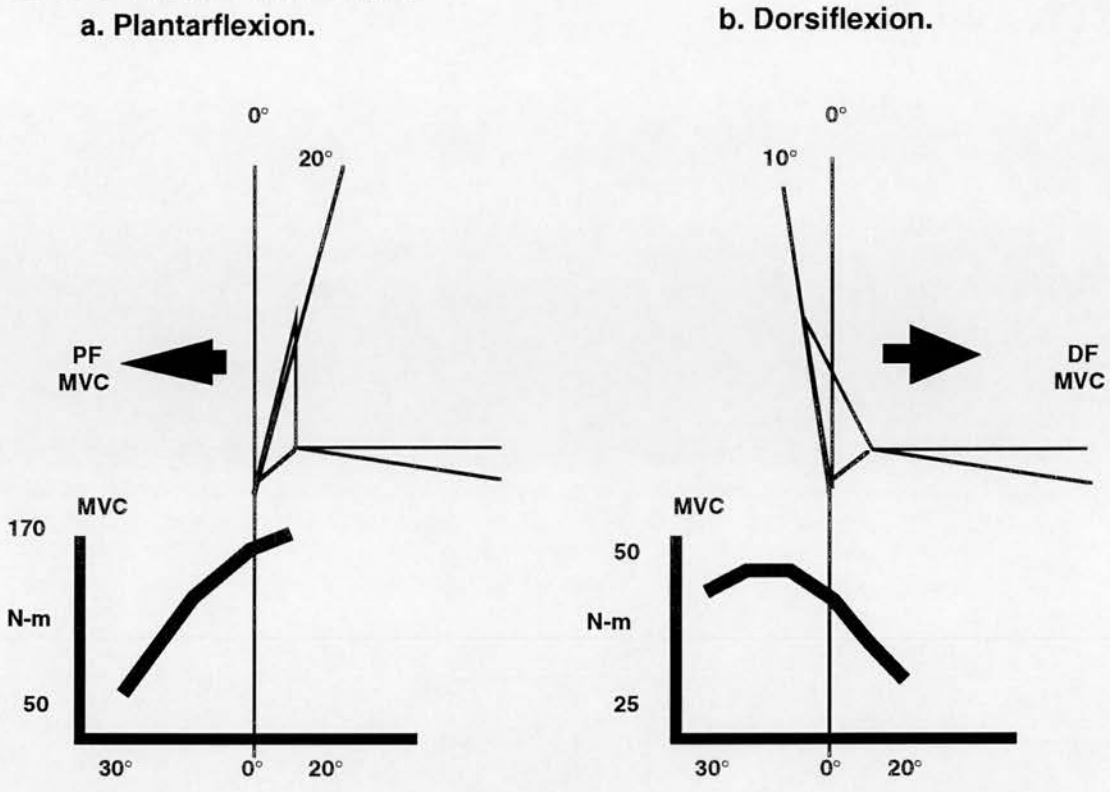


Figure 3.4.3. Mechanics at the Ankle.

Diagramatic representation of optimal maximum voluntary contraction (MVC) at the ankle joint (above) and corresponding length-tension curves (below).
a. plantarflexor power (PF) is maximal at 20° of dorsiflexion and weakest at 30° of plantarflexion.
b. dorsiflexor power (DF) is maximal at 10° of plantarflexion, rapidly decreasing beyond 5° of dorsiflexion. Note different torque scales for MVC in a. and b.
This shows that plantarflexors are weakest in equinus and that equinus favours dorsiflexion.
(After Marsh *et al* 1981; Sale *et al* 1982 in adult subjects)

After Lin and Brown, 1992, by permission.

3.4.6 Gait equinus and the hemiplegic posture.

An alternative hypothesis to explain the development of gait equinus in accord with these findings is that gait equinus results from a superimposed hemiplegic posture (see section 10: demonstration of Føg posturing in hemiplegia and tonic labyrinthine input in diplegic cerebral palsy). The hemiplegic posture is thought to emerge as a result of release of tonic labyrinthine input to the spinal cord (Magnus and Kleijn, 1912; Walshe, 1924) from a combination of pyramidal and extrapyramidal lesions of the frontal lobe, ie areas 4 and 6 (Denny-Brown, 1980). It should be stressed that gait equinus in the present study was defined for a self-selected walking speed. A different pattern of distribution of obligate toe

strikes among the 24 cases might have been observed at different speeds of walking, for instance if the children had been instructed to walk as fast as possible or to slow down.

This is an important consideration since running or sprinting, in normal individuals, appears to result in what could pass for a *forme fruste* of a physiological alternating double hemiplegia posture with bilateral alternating toe striking, hip adduction and greater extension at the knee throughout the running cycle than is normally seen in the gait cycle. This can be easily verified by sprinting on a beach and observing the single line of forefoot-prints, as opposed to heel-prints. This clearly suggests that a hemiplegic engram exists normally as an embedded sub-routine, to emerge either intermittently when required physiologically as a form of physiological decortication for running, or in varying degrees of permanence following loss of normal cortical inhibition e.g. after a stroke in childhood.

Figure 3.4.6a-b illustrates the accentuation of a hemiplegic posture in a boy with left hemiplegia (case 18): a posture which looks abnormal because of the pronated and flexed wrist and the splaying of the fingers (also seen on the good side) which are also extended as part of an involuntary Føg's posturing (Føg and Føg, 1963; see also section 10). By contrast, figure 3.4.6c-d shows a normal running posture in a girl who's upper limb posture had been abnormal in walking (ie arm extended at the elbow and pronated at the wrist) but appears normal when running.

The extent to which the hemiplegic posture is expressed must depend on the nature of the insult but at a much subtler level than that which can be analysed by the gross macroscopic lesions demonstrated by CT imaging. The often mild postural expression of hemiplegia in childhood can be unmasked by getting the child to walk, run or perform the Føg test. The threshold for the hemiplegic posture varies between resting and active states.

The hemiplegic posture depends on the child's position in space, is usually reversed upside down and is not maintained by antigravity hypertonus, muscle imbalance or spasticity as witnessed by the fact that it persists after dorsal rhizotomy (Denny-Brown, 1980). In other words, the hemiposture depends not on afferent input from the limbs, but on labyrinthine input to the limbs. Such a labyrinthine influence is clearly demonstrated in diplegic children who exhibit the phenomenon of 'scissoring' (see section 10, for demonstration).

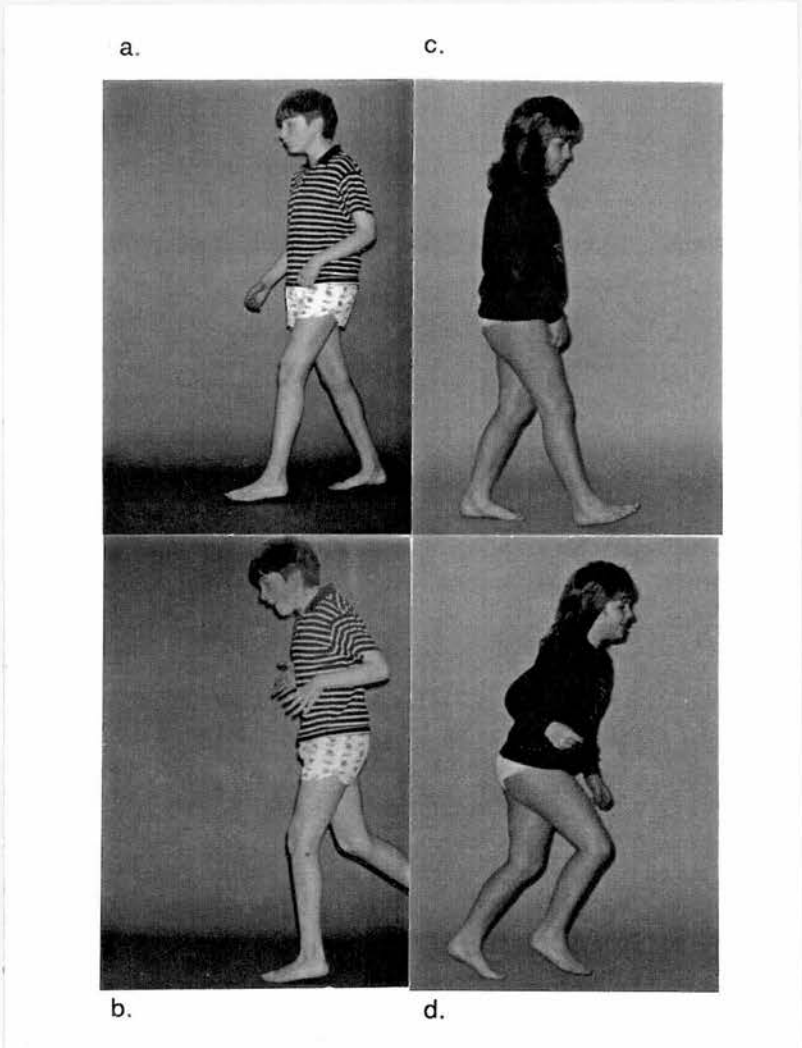


Figure 3.4.6a-d. Walking and running postures in hemiplegia.

From Lin and Brown, 1992, by permission.

3.4.7 Gait equinus: an abnormal central pattern of walking?

In a recent kinematic and EMG study of the transition from supported to unsupported walking in normal and cerebral palsied children, Leonard, Hirschfeld and Forssberg (1991), have shown that there is a common pattern of supported walking characterised by co-contraction of agonist and antagonist muscle groups (? physiological dystonia) which is normally lost as gait matures towards unsupported walking but is retained to a greater or lesser degree in children with cerebral palsy (? a prolonged physiological dystonia). Also clearly demonstrated in their kinematic analysis of ankle joint movements is the development of a passive plantar flexion following the heel-strike of the mature gait pattern (see also Sutherland, 1966 and Sutherland *et al*, 1988, pp93-97) which is absent or even prematurely activated in the independently walking cerebral palsy group.

In general the pattern of inter-joint temporal sequencing was different between supported and independent walkers with the latter, producing a distal to proximal joint activation pattern. A distal to proximal pattern of muscle activation is needed both for stability when standing (Nashner, 1985) and for ground clearance during the swing phase, and it is this distal to proximal muscle sequencing which is lost in hemiplegia as well as in the diplegias. Another important finding was of short-latency, 20-30ms segmented reflexes, most commonly originating in the lateral gastrocnemius and triggered by foot contact in the supported and unsupported cerebral palsy group.

A number of studies have examined the issue of co-contraction (persistent antagonist activation) in cerebral palsy (Milner-Brown, 1979) and this differs from observations in adults, since co-contraction was observed in children with diplegia but not adults with onset spasticity (Dietz and Berger, 1983). Gottlieb *et a*l (1982) and Myklebust *et al* (1982) advanced the hypothesis of reciprocal excitation and a loss of reciprocal inhibition to explain the findings of antagonist discharges following tendon jerks or passive dorsiflexion of the triceps muscles in cerebral palsy: a finding also found in the developing infant (Myklebust *et al* ,1986, and reviewed by Myklebust, 1990). Such a finding was greatly reduced in normal children by about 5 years of age (Leonard *et al*, 1991; Leonard and Hirschfeld , 1995).

Reciprocal inhibition of soleus motor output can be demonstrated during walking and voluntary tonic activity in healthy adults (Capaday, Cody and Stein, 1990). Using H-reflex techniques to reflexly stimulate the Gastrocnemius-Soleus muscle during voluntary

dorsiflexion, Leonard *et al* (1990) found evidence supporting a lack, or a reduced, reciprocal inhibition in children (age 6-12 years) with cerebral palsy: postulating a persistence of exuberant afferent projections. However, Berbrayer and Ashby (1990) confirmed the presence of normal reciprocal inhibition in 15 cases of cerebral palsy.

An alternative explanation for the co-contraction between agonist and antagonist pairs of muscles in cerebral palsy is the possibility of abnormal or persistent corticospinal projections to spinal motorneurones. Brouwer and Ashby (1991), using transcranial magnetic stimulation, found that in healthy individuals, only the tibialis anterior muscles can be transcranially activated, but in subjects with cerebral palsy, transcranial stimulation caused activation of *both* the tibialis anterior and the soleus muscles: supporting the notion of abnormal or persistent corticospinal projections, a finding further supported by Brouwer and Smits (1996). These studies, once again indicate mechanisms for abnormal descending motor control, independent of conventional reflex mechanisms .

3.4.8 Hemiplegic side, hemiatrophy and cerebral plasticity.

There was no bias of gait equinus or passive equinus patterns with the side of hemiplegia. Nor was limb atrophy more likely with lesions of one or other cerebral hemisphere. Botez (1971) found evidence of muscle atrophy with lesions of the post-central gyrus in the non-dominant hemispheres of patients with cerebral tumours, though he was unable to confirm a similar laterality in adult stroke patients. Alternative mechanisms of muscle atrophy in upper motor neurone lesions include secondary peripheral neuropathies, resulting from pressure to nerves (Namba *et al* ,1971) and possible "transynaptic degeneration" as a means of alpha-motor neurone loss (McComas *et al* ,1973): these two alternative mechanisms shift the "atrophy locus" away from the brain and into the periphery.

The study by Wiklund and Uvebrant (1991) could find no specific patterns of cerebral morphology associated with lower limb abnormality in their 111 cases of hemiplegia. The lack of a lateral specificity for hemiatrophy in children may testify to the plasticity of the immature nervous system injured before the development of cerebral dominance. The varying degree of hemiatrophy with muscle wasting and vasomotor instability may be in part determined by variable ipsilateral contribution from the undamaged hemisphere. Another possible mechanism protecting the limb from undergoing atrophy is the evidence for the branching of fast-conducting corticospinal pathways providing possible bilateral pre-synaptic input to the

lower motorneurone pool as a plastic response of the immature nervous system to injury (Farmer *et al*, 1990; 1991; Cohen *et al*, 1991; Carr *et al* 1991; 1993) .

There is a close similarity between the vasomotor instability and trophic changes of childhood hemiplegia documented in a previous study (Brown *et al*, 1991) and the reflex sympathetic dystrophy syndrome (RSD), with the one major exception that the hemiplegic limb is rarely, if ever painful. Whether these disparate phenomena share a common central mechanism involving abnormalities of sympathetic control or of peripheral opioids (Hannington-Kiff, 1991) may emerge in due course.

3.4.9 Motor dexterity.

There was no clear association between reduced fine motor dexterity and gait equinus or passive equinus. This finding was expected since fine motor dexterity was universally reduced for all the affected limbs and could scarcely account for differences between one hemiplegia and another.

3.4.10 Passive equinus and peripheral variables.

In contrast to gait equinus, there was a positive association between passive equinus (reduced compliance) and a shorter leg, reduced ankle joint range, reduced extensibilité and increased resistance to passive stretch corresponding to a clinical impression of 'tonic spasticity'. Sharrard (1964) analysed the relationship between the degree of 'spasticity', graded as mild, moderate or severe, and equinus deformity in 104 spastic limbs but was unable to find any correlation between the two. His clinical assessment of spasticity was confined to eliciting phasic spasticity with rapid dorsiflexion, making no comment on the presence or absence of tonic spasticity. A re-examination of Sharrard's data on equinus deformity and phasic spasticity with chi-square analysis appears to show a highly statistically significant association between moderate to severe phasic spasticity and equinus deformity. However, a further analysis of splitting the chi-square table suggests that equinus deformity is unlikely to develop with mild phasic spasticity, whereas an increasing severity of phasic spasticity and equinus deformity are not statistically significantly associated.

Two thirds of the present 24 hemiplegic children had evidence of reduced calf muscle compliance. Although a more detailed analysis of the muscle tone and reflex excitability is to follow, it was not possible to correlate resistance to passive stretch at rest with EMG activity, i.e. there did not appear to be EMG activity at the time of tonic muscle

stretching. These findings suggest a peripheral biomechanical transformation of muscle rather than spasticity as a cause of the resistance to passive movement at the ankle joint in childhood hemiplegia (fig. 3.4.4)

3.4.11 Implications for management of equinus in hemiplegia.

The careful clinical assessment can go some way to identifying the group determinants of abnormal waling patterns but more diagnostic information, capable of advising the clinician and therapist is required. An apparently mandatory addition arising from this study is the need for detailed information about the activity of the muscles as an aid to decision-making: clearly a minimum amount of information is whether the muscles are 'on' or 'off'; whether the muscles interplay or co-contract; whether they respond abnormally to stretch in a phasic or tonic manner and so on.

The evidence so far suggests that an equinus gait is multifactorial. As indicated in figure 3.4.2, 'treatment' could be directed at different levels. For example, head position could result in maladaptive posturing (flexion of the arm at the elbow, adduction and internal rotation of the leg at the hip, extension at the knee and equinus), spinal cord disinhibition might warrant treatment by a variety of methods aimed at reducing overactive reflexes and electrical tone; peripheral transformation of muscles would require a different group of treatments, each of which need to be individually tailored to suit the child. As will be seen the relative contribution of each of these mechanisms can be assessed.

The current evidence suggests that the principal causes of gait equinus are

1. The expression of the hemiplegic posture to varying degrees.
2. A disturbance in central motor activation patterns.
3. Biomechanical (rheological) changes within the muscles.
4. Abnormal phasic activation of the plantarflexors after foot-contact or in pre-positioning of the foot in terminal swing

Muscle strength and muscle tone do not appear to contribute directly to gait equinus. The classical concepts of pathologically strong, spastic, shortened calf muscles and weakened, elongated anterior tibial muscles producing equinus is at variance with what can be demonstrated about the MVC about the ankle joint since an equinus posture, if anything, favours dorsiflexor power. Limb dwarfing, reduced ankle joint range and diminished extensibilité are not causes of gait equinus but consequences of the same cerebral injury

producing gait equinus. These peripheral changes are most likely to affect the efficiency of walking and running, reducing speed and increasing the energy cost of such activities.

Splinting the ankle with an ankle-foot orthosis (AFO) corrects the central disturbances of a hemiplegic posture, an abnormal walking engram and neutralises the "kick" produced by the spinally-mediated phasic reflex excitability and may improve the efficiency of walking. The AFO and plaster cast may operate by chronically loading the calf muscles (see section 7). A tendo-Achilles tenotomy or a muscle recession procedure, may, by contrast, result in chronic unloading and weakening of the already weakened calf muscles (see section 7). Tenotomy completely neutralises all central, spinal and peripheral contributions to hindfoot equinus since the Achilles tendon is the final common path the muscle effector organ. Tenotomy acts also as a form of deafferentation procedure since the slack extrafusal muscle fibres result in slack intrafusal fibres (Vrbová, 1963). The two main treatments available for equinus: stretching and releasing the muscle, may have opposite physiological effects on the muscle.

3.5 Summary of findings.

i. A detailed clinical examination of 24 hemiplegic children at a mean age of 9.9 years (SD 3.0) showed that 9/24 had an obligate toe strike when walking at a self-selected speed and this was maintained throughout stance.

ii None of the 24 children had a fixed equinus.

iii. No association could be found between this pattern of gait equinus and lower limb atrophy, reduced ankle joint range, muscle extensibilité, power of dorsiflexors and plantarflexors or actual muscle imbalance at the ankle joint.

iv. Gait equinus was independent of reduced compliance of the calf muscles or a clinical diagnosis of tonic spasticity or of fine motor dexterity of the toes or the side of the hemiplegia.

v. Gait equinus cannot be explained merely in terms of a central paralytic foot drop.

vi. A parallel finding of reduced compliance or passive equinus, independent of gait equinus, was associated with the most dwarfed limbs, those with the narrowest joint ranges and greatest differences in muscle extensibilité together with clinical evidence of tonic spasticity.

vii. Passive equinus was independent of power ranges, motor dexterity or the side of

hemiplegia.

viii A developmental model of equinus in keeping with these findings is advanced.

3.6 Future areas for research.

Relatively scant attention has been paid to the pathological or adaptive changes of muscle in cerebral palsy. How much is disuse or misuse or susceptible to therapy is impossible to say without further study of muscle transformation in selected groups. There are therefore four important areas of enquiry:

Motor development.

Peripheral muscle transformation in cerebral palsy.

Reflex excitability and abnormal muscle contraction.

Mechanics at the ankle joint.

3.7 Acknowledgements.

Part of this study has been published elsewhere by Lin and Brown, 1992, (seeAppendix).

4. Plastic Properties of Muscle in Hemiplegic Cerebral Palsy.

4.1 Background.

The clinical evidence presented in section 3 suggested that the muscles of hemiplegic children offered a non-neural resistance to stretch: a stretch not accompanied by or resisted by concomitant electrical activation of muscle. The clinical importance of this phenomenon, if borne out, would be important because of its implications for management in such cases which, at the very least would exclude pharmacological and neurosurgical attempts at reducing muscle tone.

Foley (1960) indicated in his studies in diplegia, that stretched muscles behaved in a way which indicated rheological change. This was also reported by Herman (1970), who noted that the muscles of his adult hemiplegic patients did not immediately return to their resting length after being stretched. This was particularly the case in a group of patients in whom reflex excitability was low and in which clonus was not elicitable, and Herman (1970) classed these limbs into a pre-contracture group (Group IV). This contrasted with other limbs which were respectively: atonic-flaccid (group I), limbs which possessed weak reflexes with little or no clonus (groupII) and limbs with strong reflexes and marked clonus (group III). The degree of reflex excitability and non-excitable muscle resistance appearing to vary with the age of the injury.

Dietz, Quintern and Berger (1981) presented evidence to suggest that in adults with chronic spasticity, altered mechanical properties of muscle contribute to hypertonia during the walking cycle. In a further study involving 30 healthy controls (20 adults and 10 children) 54 patients with hemiparesis, 24 with a paraparesis and 10 children with cerebral palsy (6 with diplegia), Dietz and Berger (1983) found that:

> "The surprising result of our experiments was that there is no electrophysiological explanation for leg muscle hypertonia. We suggest that muscle stiffness during locomotion in spastic patients is due to changed mechanical properties of muscle fibres."
>
> *Dietz and Berger, 1983.*

Hufschmidt and Mauritz (1985) demonstrated a chronic peripheral transformation of muscles of adults suffering from a variety of neurological diseases such as stroke, hemispheric glioma, cervical myelopathy, multiple sclerosis, tetraparesis, paraplegia and hereditary spastic paraplegia: the duration of the injury ranging from 6 weeks to 22 years. These authors showed that the work absorbed during slow muscle stretches ie stretched at

rates between 2°/s and 20°/s, was increased in the patient group. These slow speeds of stretch did not excite any reflex responses whereas velocities of stretch between 80°/s and 200°/s produced increases in the work of stretch attributable to reflex activity. In their study, Hufschmidt and Mauritz (1985), did not observe electromyographic activity at rest, nor was there any evidence of a tonic stretch reflex response in their patients. To make doubly sure, Hufschmidt and Mauritz (1985) abolished Ia afferent input by applying an ischaemic block, following which the work of stretching the muscles was unchanged. Such a viscoelastic transformation was apparent in patients whose illness exceeded a year's duration; the work of stretch increasing with the time elapsed after the stroke. The biomechanical component supporting the resistance was attributed to plastic muscle change rather than viscous change, since the authors were not able to demonstrate a viscous component at the slow speeds of stretch (2°/s-20°/s): the resistance encountered being referred to as 'plastic'.

> "In contrast with other workers, we found that the amount of work absorbed was not significantly related to angular velocity (2° to 20°/s). As viscosity is positively rate-dependent by definition, this finding demonstrates that at physiological rates of movement (20°/s corresponds to normal walking) human lower leg muscles exhibit no measurable degree of viscosity. This is also true in spastic patients. The resistance encountered in unexcited muscle beyond short-range elasticity should therefore be termed "plastic".
>
> *Hufschmidt and Mauritz , 1985.*

The same authors were able to show an increase in the passive elastic stiffness in Parkinsonian subjects (1991) without any changes in the contractile properties of the Parkinsonian muscles.

These phenomena of non-electrical hypertonus or resistance to stretch are similar to the 'passive equinus' determined by clinical examination in the 24 hemiplegic children presented in section 3 above, but clearly there are implications for children with bilateral involvement since 6/10 of the Dietz and Berger (1983) children had diplegia.

The following studies illustrate the phenomena of plastic muscle change, namely: 'creep', 'stress-relaxation' and 'moulding' in a few cases of congenital hemiplegia. One object of these studies was to define the clinical characteristics of children whose muscles exhibit plastic change.

4.2 Methods.

4.2.1 Manual stretches

Manual stretches were applied as in section 3 above but in addition, the joint angle was monitored at the ankle using a Penny and Giles Twin Axis Flexible Goniometer attached

to back of the calf and the heel. The goniometer was 'zeroed with the sole at 90° to the shaft of the tibia: ie the neutral angle.

4.2.2 Instrumented torque stretches

A pre-set trapezoid torque was delivered by a large printed motor (MC24P, torque constant 41.5Ncm/amp) which was mounted coaxial to the ankle joint and an optical potentiometer, with the subject side -lying and the leg extended. The foot rested on a pedal attached to the lever arm.

Surface electromyographic (EMG) activity was recorded from the tibialis anterior and triceps muscles. The individual experimental conditions are explained for each case presented.

4.3 1 Results.

Case one is a 14 year old boy with a congenital left hemiplegia. The left hemiatrophy is evident in figure 4.3.1 (same child as in figure 3.4.6a-b, case 18): which shows a thin left arm and left leg, with a characteristic left hemiposture which was noted to vary according to the level of activity and rest. The foot contact pattern is that of a toe-strike, so-called equinus gait, instead of the normal mature heel-strike which occurred on the non-paretic side.

4.3.2 Is this muscular creep?

Figure 4.3.2 illustrates the changes in joint angle during passive manual dorsiflexion of the foot at the ankle along with simultaneous surface EMG recordings. The top traces relate to the nonparetic limb, the lower traces to the hemiparetic limb of the child in figure 4.3.1. For both legs, the Tibialis anterior (TA) and Gastrocnemius-Soleus (G-S) muscle EMG has been recorded in volts (V) with surface electrodes and the ankle joint angle recorded in degrees (°) with a potentiometer concentric with the ankle joint. The time in seconds is shown on the horizontal axis. It is important to note that 0° corresponds to the right angle position of the foot with the shaft of the tibia. For both the nonparetic and hemiparetic limbs, the initial joint angle corresponds to the *resting angle* .

When the examiner passively stretches the resting nonparetic calf, little resistance is met and the muscle can be stretched easily across the full joint range a number of times in a short space of time (about 15 s). The subject is completely at rest, and the EMG is silent on the nonparetic side apart from two bursts of EMG prior to the onset of the stretch manoeuvre and a 0.5s burst in both muscle groups (TA and G-S) at maximum dorsiflexion during the third

Figure 4.3.1. Congenital left hemiparesis with gait equinus.
(Case 18 section 3, see also figs 3.4.6a-b)
For explanation, see text.

stretch. The rate of stretch is uniform for all four stretches. There is little change in the resting joint angle at the end of the series of stretches when compared with the initial resting angle. The examiner exerts little effort in performing these stretches.

On the hemiparetic side (bottom trace), the resting angle is twice as plantarflexed as on the nonparetic side and the examiner encounters marked resistance to stretch so that he is obliged to apply a large manual torque to achieve full dorsiflexion. But if he maintains a steady dorsiflexing torque, the muscle gradually plays out and appears to *creep* slowly at a constant perceived tension towards it's final length. Note that the slope of the change in angle appears to have four phases: phase 1 lasts about 0.6s and is the steepest, phase 2 lasts 3s and phase 3 lasts 8s from 6s to 16s: thereafter, phase 4, the angle is maintained constant for 1s before the manual torque is released. The full joint range is achieved very slowly, in this case over 14s instead of 2.5s on the nonparetic side.

Throughout this manoeuvre, the hemiparetic muscles are electrically silent: the resistance offered is therefore not reflex or supraspinal in origin. Clearly, the muscle on the hemiparetic side has changed compared with the nonparetic muscle.

4.3.3 Muscle "Moulding".

Figure 4.3.3 indicates the change in resting plantarflexion after four sequential manual dorsiflexion stretches applied to the hemiparetic limb of a 7 year old boy with a congenital hemiparesis. The time is measured in seconds and the joint angle in degrees. Note how following each stretch, the resting joint angle has changed by moving closer to the neutral or 0° position. This is achieved after the fourth and final stretch, some 80 seconds after the first stretch. The muscular creep occurs after the initial rapid and virtually linear dorsiflexion. The total creep over the four stretches amounts to 53°. After release, the muscle does not return to its original length: the cumulative new length is most apparent after the fourth stretch, the new resting angle is temporarily -7° instead of -60° at the outset. This sustained effect of preceding stretches can be viewed as a form of muscle moulding, much as a piece of plasticine or putty can be moulded or deformed (see section 2: plastic properties of muscle).

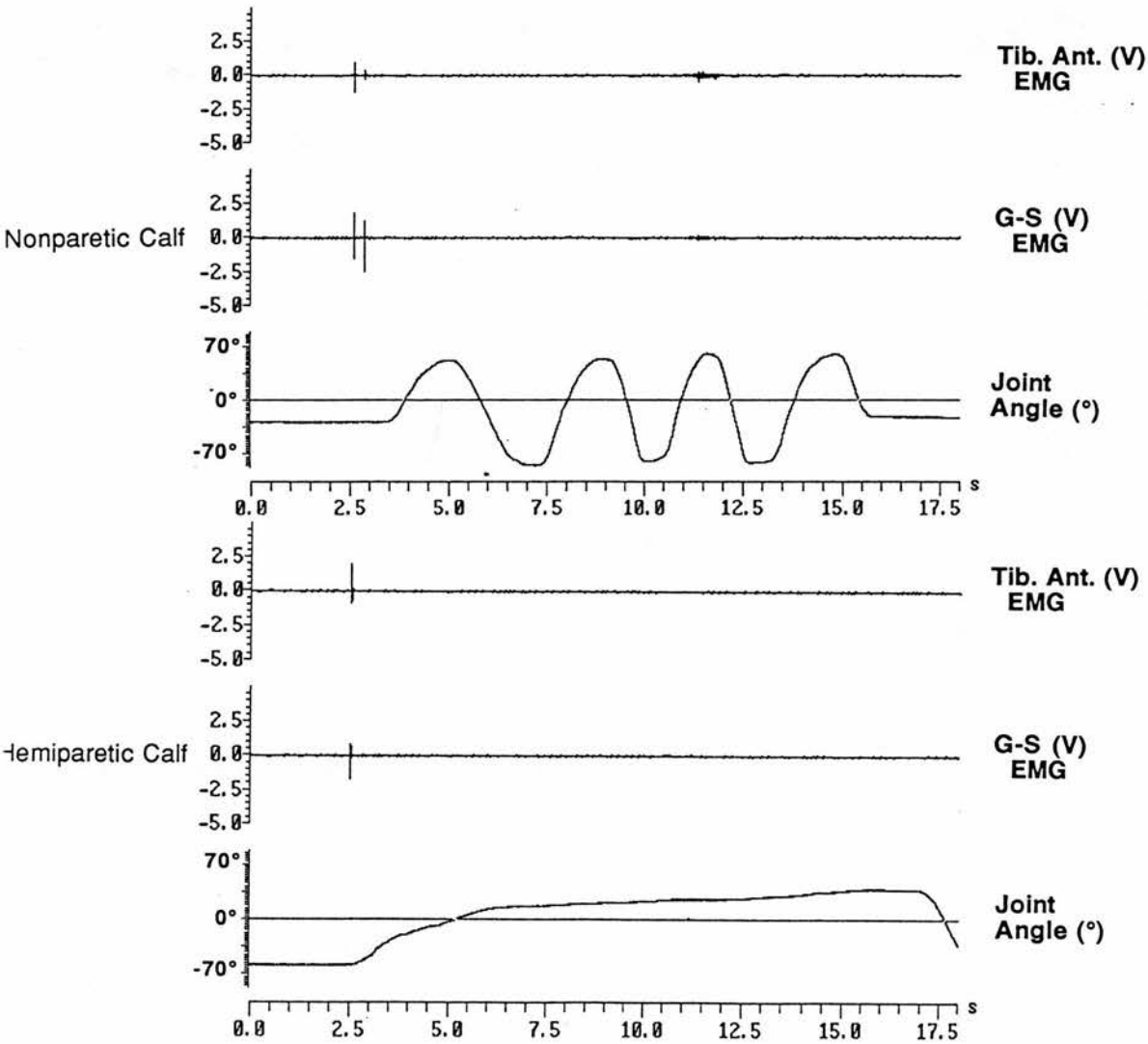


Figure 4.3.2 Manual dorsiflexion across the joint range (for explanation see text) (Case 18 section 3, see also figs 3.4.6a-b, and 4.3.1). For explanation see text.

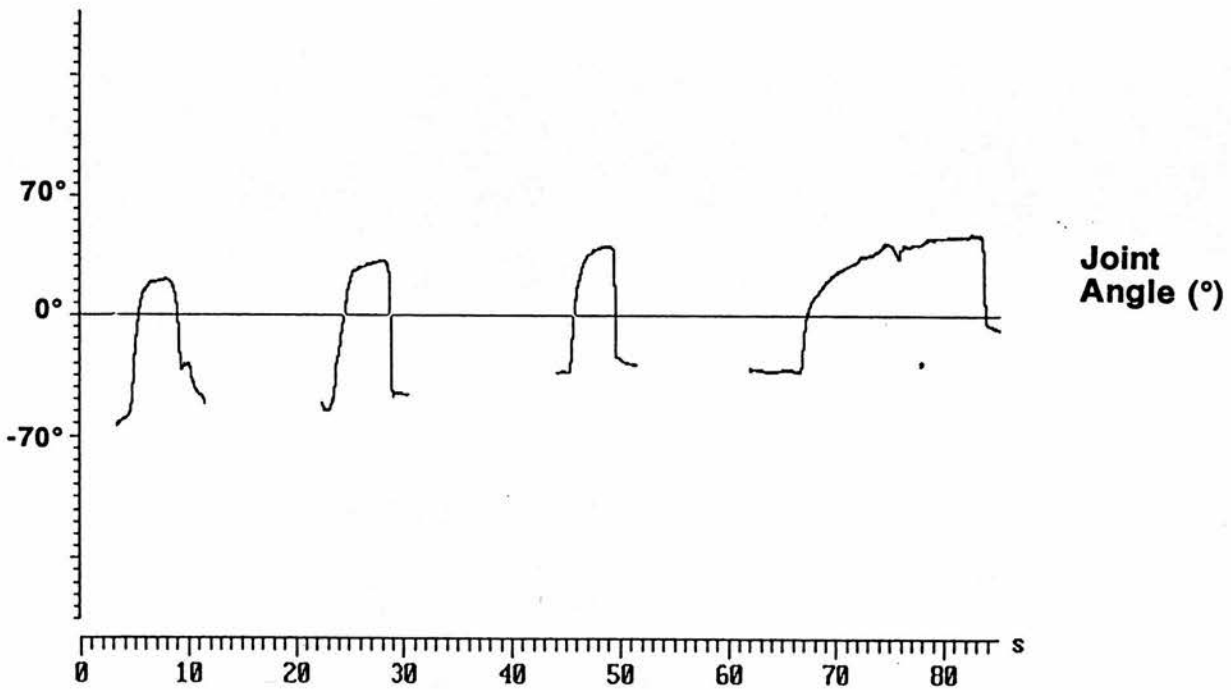


Figure 4.3.3 Muscular creep demonstrated after four sequential stretches.
7 year old boy with congenital hemiparesis. For explanation see text.

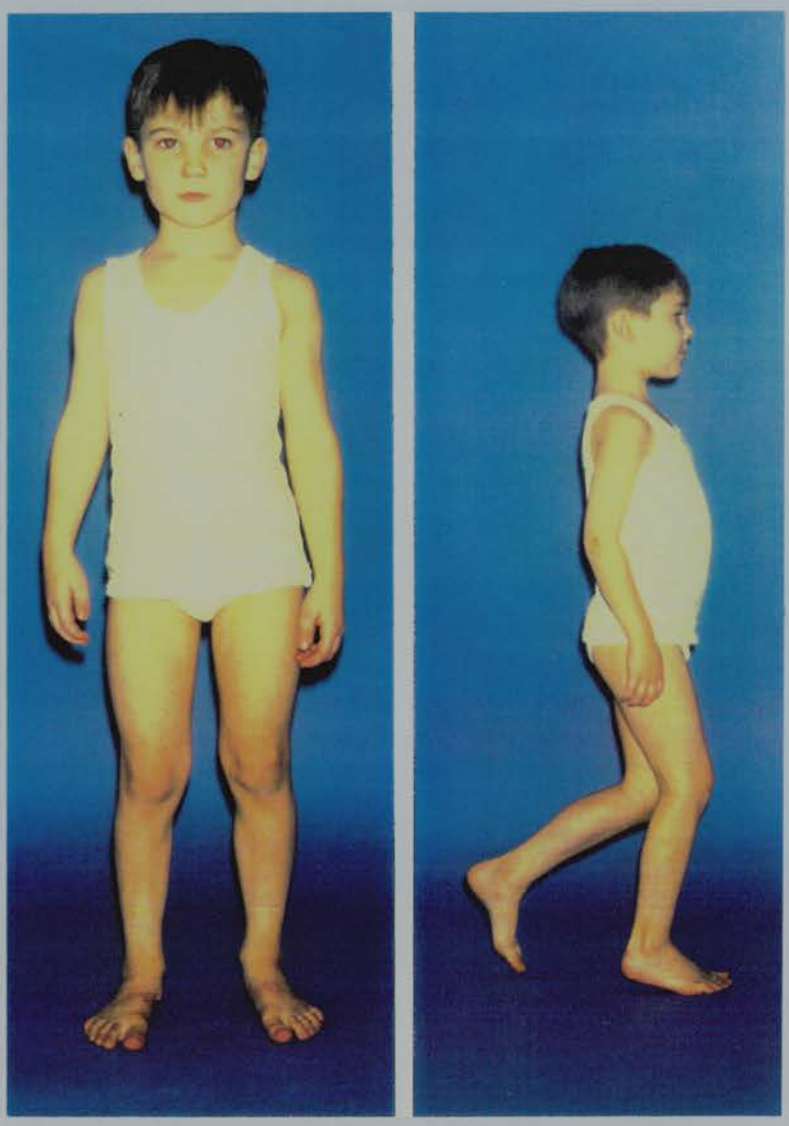


Figure 4.3.4.i Eight year old boy with congenital right hemiparesis. (case 17, see fig. 3.3.1). Showing equinus gait.Note that right calf muscle bulk is relatively well-preserved with little limb asymmetry..

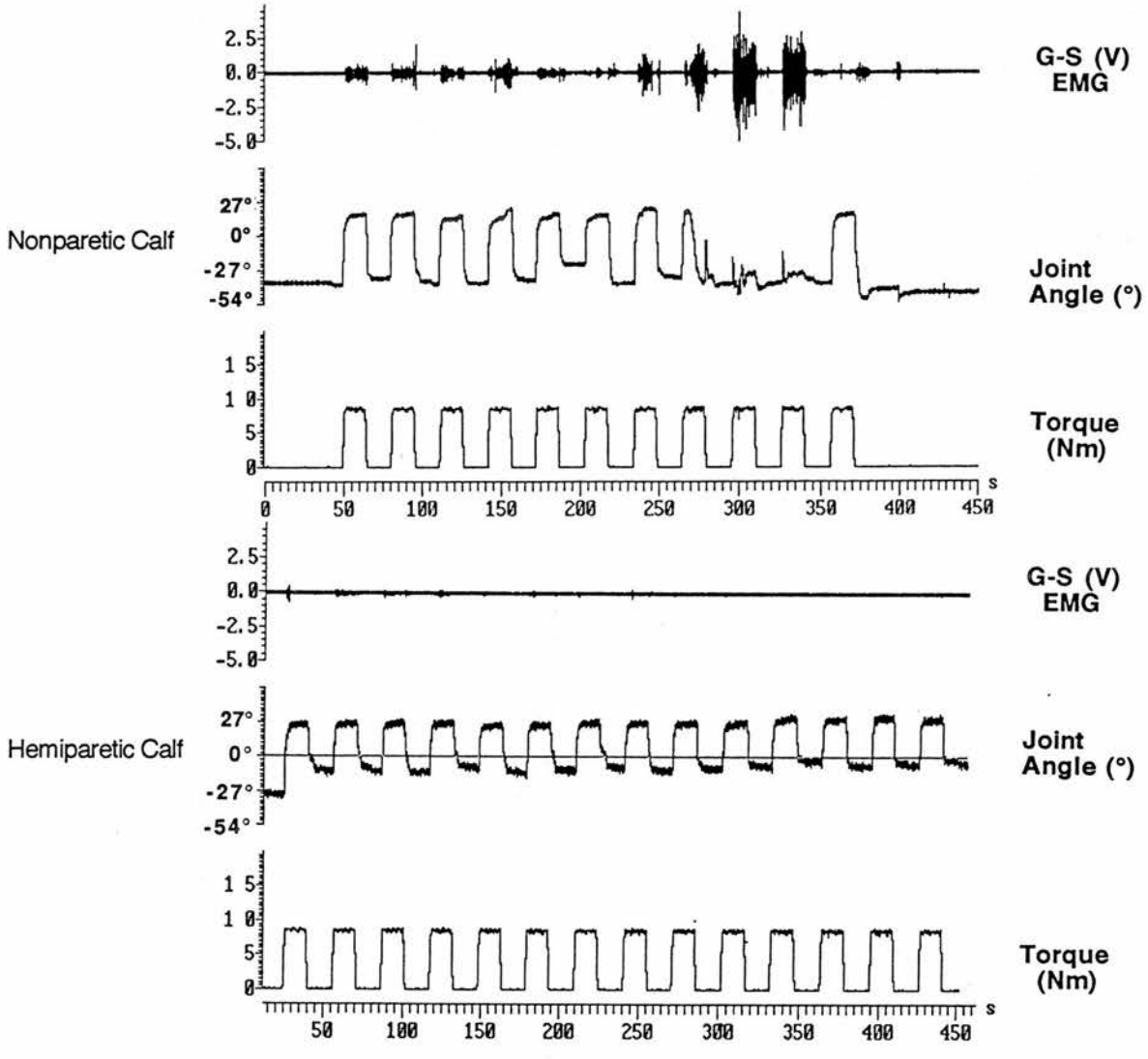


Figure 4.3.4.ii Eight year old boy with congenital right hemiparesis.
Torque motor stretching of soleus muscles with knee flexed at 90°. Top: nonparetic side showing voluntary resistance to passive stretch. Bottom: hemiparetic limb shows muscular creep. Middle trace of each panel: 0°= neutral angle, upward deflections=dorsiflexion (°). Lower trace of each panel: upward deflection=dorsiflexor torque (Nm).
For additional explanation see text.

4.3.4 Instrumented creep.

So far, clinical examples have been shown to illustrate creep. One difficulty is not knowing whether the examiner is able to maintain a constant dorsiflexing torque. A study was performed using a large printed motor to apply 10Nm dorsiflexing ramp torques which were then sustained over 13 seconds. This study involved an 8 year old boy with a right congenital hemiparesis (fig. 4.3.4.i). In this example (4.3.4.ii), the nonparetic limb is shown above the hemiparetic limb. It is noteworthy that EMG activity occurs on the normal left side which is tonically resisting the attempted dorsiflexion. In fact, the subject easily neutralises the dorsiflexing torque pulses on the good side between 275 and 350s into the study, and the constant competition between the subject and the motor causes the resting joint angle to 'wander' erratically.

On the hemiparetic side, there is little or no EMG activity in response to a succession of 10 Nm torque pulses. The initial pulse stretch results in a dramatic alteration in the resting angle which is more than halved. After the tenth pulse, the resting joint angle between pulses, gradually moves towards the neutral joint angle, although the first stretch produces the maximum creep and the maximum change in the resting angle.

4.3.5 Reproducibility of muscle creep and duration of "moulding".

The traces in figure 4.3.5 represent the triceps EMG, joint angle and torque applied to the hemiparetic limb of the 14 year old boy (case 18) who had been examined clinically in section 4.3.1-2 above.

After an initial dorsiflexion at an average angular velocity of 22°/s under a slowly rising torque lasting 1.07s, the muscular creep is clearly evident, as a rounded "shoulder"on the angular trace, during the constant phase of the 10 Nm torque, the joint angle creeping up to a constant angle over 7.18s at an average velocity of 1.27°/s. Notice how the resting joint angle does not return to its original position when the dorsiflexing torque ceases.

This moulding of the muscle to a longer resting length persists for 80 seconds during which there is a gradual non-electrical shortening towards the original resting length. This is punctuated by two relatively abrupt or rapid small plantarflexions which are not accompanied by any triceps EMG. When the muscle has been rested for 22 seconds, a second torque pulse is applied and maintained at 10Nm with similar effect.

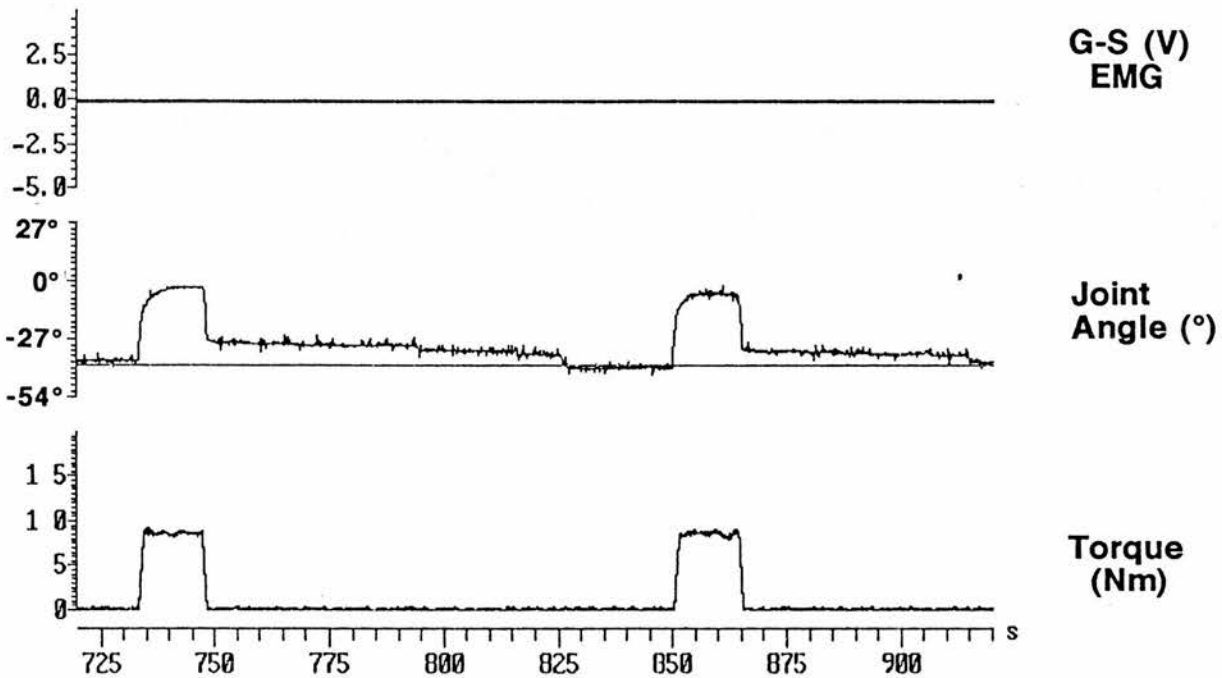


Figure 4.3.5 Demonstration of muscular creep and moulding.
Case 18: see figs 4.3.1 and 4.3.2
For explanation see text above and below.

Again creep is apparent with a corresponding shift in the resting joint angle closer to the neutral angle, ie muscle moulding to a longer length. The muscle gradually shortens again over the next 126 seconds: the plastic change in the muscle length thus obtains over several minutes.

A particularly important feature of muscle creep is the fact that the new joint angle or muscle length can be maintained with a diminishing dorsiflexing torque: such a phenomenon has been referred to as *stress-relaxation* (see Foley, 1960, Wright and Johns, 1960). Presumably, this is what enables an ankle to be held by an orthosis in marked contrast to a dystonic limb in which strong muscular contractions accompanied by excessive EMG activity rebel against the splint or the examiner. Such limbs are impossible to fit into an orthosis since the tension in the calf muscles is always extremely high.

4.3.6 Summary.

i. A clinical and instrumented demonstration of muscular creep has been presented indicating a peripheral transformation of the plastic or time-dependent properties of muscle following long-standing cerebral injury which, in the case of these few children, means life-long.

ii. Two further corollaries of muscle creep are:

a. Muscle moulding or short-term adaptation to a longer resting length which persists for several minutes or more if the dorsiflexing torque is removed.

b. Stress-relaxation which allows any given hemiparetic muscle length to be maintained with a diminishing dorsiflexing torque.

iii. Muscle creep is independent of EMG activity, ie is non-reflex and not dependent on supraspinal electrical 'tonus'. It is predicted that creep would be present in the anaesthetised / curarised subject and would not be affected by such procedures as selective dorsal rhizotomy, intramuscular botulinum toxin A injections or more conventional muscle relaxants such as baclofen (acting on pre-synaptic GABA B receptors) or diazepam(acting centrally on GABA A receptors).

iv. Physical therapies such as repeated muscle stretches, the use of physical exercises, orthotics and serial plastering, presumably depend in part on the property of muscular creep and stress-relaxation for their success: ten to twenty seconds of stretch may allow day-long wearing of an orthosis or plaster-cast at the ankle without undue tension in the calf muscles. This is in contrast to the dystonic limb or the limb under the influence of strong labyrinthine or tonic neck reflexes that will 'fight' the splint, producing pressure sores. (see section 3 and 9)

v. The natural history of muscular creep in the congenital hemiplegias is not known. However, procedures aimed at lengthening the muscle-tendon complex must take into account the presence of muscle creep to avoid the disastrous consequences of over-lengthening and sinking (or creeping) into crouch.

5. Velocity-dependent Proximal Lower Limb Reflex Excitability.

5.1 Background.

An examination of the current therapies available for the management of the cerebral-palsied child testifies to the optimism and/or desperation of the carers. These include physical therapies, orthotics, plaster immobilisation, drugs (oral and intrathecal), tendon and bone surgery (alone or in multiple combinations), local botulinum toxin injections to the muscle, nerve blocks and finally selective dorsal rhizotomy (SDR). But the advent of invasive treatments such as intrathecal baclofen and the recent rehabilitation of Foerster's rhizotomy in the form of selective posterior rhizotomy has made it necessary to review the basic assumptions about "the spasticities" (see sections 1.3, 1.5 and 2.3.5).

Essential to any argument about the benefits of one treatment over another is a clear definition of what spasticity is or is not (Landau 1974). The functional significance of spasticity as the result of damage to the growing and developing brain continues to be controversial. The patterns of disturbed tone arising from damage to the adult brain may differ from those of the child with cerebral palsy: indeed recent evidence from work with children suggests that new pathways are laid (or their regression offset) by early injury to the motor system (Carr *et al*, 1991; Cohen 1991).

There is a prevalent assumption that abnormal stretch reflexes are major contributors to the movement disorder in cerebral palsy. The elicited stretch reflex, along with voluntary and involuntary muscle activity, combine with the biomechanical properties of the muscle-tendon-joint complex to offer resistance to stretch (see section 2 for a full discussion).

One obvious shortcoming of the many clinical outcome studies has been a failure to grasp that cerebral palsy syndromes comprise a disorder of *movement*, of *posture* and of *muscle tone*. The disturbance of tone may be due to *tonic labyrinthine release*, excessive expression of the *tonic neck reflexes*; it may be due to excessive *Føg's posturing*,*dystonia*, *spasticity* or related to intrinsic *biomechanical* changes in the properties of the muscles ie both the well-understood viscoelastic and the less well-known *plastic* properties of muscle.

As discussed in section 1.3, 1.5 and 2.3.5 of this work, the term spasticity has been used in at least two different ways. On the one hand, the term 'spasticity' is used to describe a clinical syndrome (see Crothers and Paine, 1959 and section 1.3 above) which attempts to encompass the totality of disordered motor manifestations, including the disordered

movements and postures. This contrasts with an operational definition of spasticity relating the phenomenon to a quantifiable physiological response to a set of well-defined stimuli.

This dichotomy is brought into stark focus if one considers the implications of using the syndromic terminology of spasticity in relation to a very specific procedure such as dorsal rhizotomy: does rhizotomy relieve the effects of supraspinal influences such as the tonic labyrinthine release? The answer according to Denny-Brown (1980) is that the hemiposture is not relieved by rhizotomy. Accordingly, either **i.** rhizotomy is not a treatment for spasticity or **ii.** the broad syndromic definition used is incorrect. Indeed, both **i.** and **ii.**are logical extensions of the the current muddle. Obviously, rhizotomy cannot be expected to 'treat' the whole motor syndrome, but it does reduce or abolish phasic phenomena such as brisk reflexes and clonus. Rhizotomy, in fact 'treats' the restricted, operationally-defined spasticity but may have little or no effect on the other, perhaps more important syndromic accompaniments.

As will be seen, to be used meaningfully, the term spasticity needs to be restricted to a subset of the total motor disorder relating to velocity-dependent stretch reflexes which are abolished by rhizotomy: The other motor accompaniments such as weakness, lack of motor dexterity, co-contraction, tonic labyrinthine release (eg hemiposture, scissored posture of diplegia), tonic neck reflexes and biomechanical transformation of the muscles over time need to be seen as coexisting disturbances, each of which are likely to need separate strategies for management. Section 3 has already described the multifactorial disturbances leading to gait equinus and section 4 has briefly demonstrated the presence of 'plastic' (time-dependent) change within muscles in hemiparetic CP. This section deals with the velocity-dependent ie 'phasic' reflex disturbances of motor control.

5.2 Ideal theory and measurement of spasticity.

The *ideal theory* of spasticity should **a)** be capable of describing the phenomenon in clear operational terms, **b)** explain why it sometimes takes days or often weeks for spasticity to emerge after spinal or cerebral injury, and finally, **c)** offer clues for the treatment and rehabilitation of the patient which are likely to be beneficial. The classical models for spasticity have involved the study of the roles of the muscle spindle (intrafusal fibres), type Ia and type II sensory nerve afferents, alpha-motorneurone, gamma-motor neurone (fusimotor) and spinal interneurones in the pathophysiology of spasticity. Such studies have established that there

is no change in muscle spindle sensitivity to stretch and no evidence for fusimotor overactivity in the production of spasticity. Studies using muscle vibration have established the likelihood of a loss of pre-synaptic inhibition between the dorsal root afferents and the alpha motorneurone resulting in a lowering in the reflex velocity threshold and an increase in reflex gain to a given velocity of muscle stretch as measured by the electromyographic (EMG) response of the muscle.

The *ideal measurement (s)* should be simple to use and relate to the pathological process as well as offer some prediction of the likely effects of intervention for planning a course of treatment. Many therapies, physical, pharmacological or surgical, are directed towards alleviating "stretch reflex hypertonus", though few studies have demonstrated a reduction in abnormal reflex activity or functional improvement after treatment.

Such a lack of corroborative evidence is due in part to the difficulty in quantifying reflexes and reflex change. The problem is further compounded by the fact that not all EMG activity recorded is reflex, some being voluntary, some involuntary but mediated by descending supraspinal mechanisms and some truly due to spinal and supraspinal stretch reflex mechanism (see section 3.4.7 for discussion).

Many studies in the past have settled for recording the clinical resistance felt on passive stretching according to the Ashworth scale (Ashworth, 1964), which was originally devised to assess spasticity in multiple sclerosis patients. The use of such a non-linear, nonparametric, ordinal scale of severity does not warrant the uncritical attribution to spasticity of the resistance felt in the subjects to whom it is applied, it merely grades the resistance felt irrespective of the factors producing such resistance, and this may have lead to a number of erroneous conclusions in the past, relating to treatment and outcome. The use of the Ashworth rating does not eliminate the need to define each component of the stretch resistance.

The most commonly accepted definition of spasticity is that of Lance,1980:

"In both cerebral and spinal spasticity, the stretch reflex responses obtainable from extensor and flexor muscle groups of the upper and lower limbs increase approximately linearly with increase in the velocity of stretch. The reflex component of the increased tone may therefore be measured in terms of the threshold velocity required to evoke reflex activity and the slope of the EMG-velocity relationship."

Lance, 1980.

It follows that the velocity threshold of stretch necessary to elicit a reflex is reduced

and the amplitude of a reflex response to a given velocity of stretch is increased in spasticity compared to the normal. A wide variability of responses is said to be the hallmark of normal reflex excitability whereas the spastic subject exhibits a stereotyped exaggeration of reflex activity which is only occasionally encountered in normal subjects (Neilson and Lance, 1978; Rack *et al* 1984).

The use of step-torque averaging techniques (equivalent to a ramp stretch manoeuvre) have demonstrated that the upper latency of a true polysynaptic reflex response occurs in less than 90 ms when actively contracting wrist flexors are perturbed (Lee and Tatton, 1978) or 150 ms for the triceps surae muscle (Berardelli et al 1982). This methodology sets a latency limit on what may be regarded as stretch reflex activity: according to this definition, any EMG activity occurring outside 90 ms for the wrist, or 150 ms for the calf, is not a stretch reflex. A large number of other phenomena may be responsible for non-stretch-reflex involuntary motor activity: for example, section 10 illustrates the role of tonic labyrinthine responses (see Chan, 1983, for a detailed discussion) and Føg posturing (Føg and Føg, 1963) and these neurological events are likely to fall outwith the latency range for the stretch reflexes in upper and lower limb.

The study of reflex behaviour requires knowledge of the starting muscle length, which corresponds to the initial joint angle together with the amplitude and velocity of stretch. The state of the muscle prior to stretch, whether resting or active and the method used to stretch the muscle should be specified. By combing a sinusoidal with a ramp technique, many aspects of velocity-dependent reflex activity can be assessed.

Traditional tendon jerks, though excellent diagnostic indicators of corticospinal damage, are difficult to grade and are produced by a stimulus which combines stretching and vibration of the muscle spindle (Lance and de Gail 1965). Tendon jerks may be considered unrepresentative of the types of motions or stimuli commonly encountered in every day life and as such, it may be thought that the applied stimulus is unphysiological, provoking a supramaximal, all or nothing, as opposed to a graded reflex response.

The following studies attempt to investigate such a graded stretch reflex quantification to graded stretches applied to the thigh muscles and section 6, describes similar studies applied to the calf muscles.

5.3 Methods.

The aim was to measure the reflex electromyogram (EMG) elicited by applied stretches, at varying speeds though the resistance felt during the stretches (which would require other techniques) was not recorded.

5.3.1 Patients.

A group of 13 children with congenital hemiplegia aged 6.2 to 12.2 years (mean 9.4), described in a previous clinical study (Lin and Brown 1992), underwent evaluation of proximal lower limb reflex excitability under electromyographic control. All had congenital hemiplegia. Perinatal details include prematurity in 2 cases, intrauterine growth retardation in 2 cases, birth asphyxia in a further 2 cases and the remaining 7 cases were born at term without known antenatal or perinatal difficulty.The 13 children involved in this study were cases 4, 6, 8, 10, 11, 12, 14, 15, 16, 17, 18, 22, and 23. Further details of the patient population are given in table Table 3.2.1, above, including a clinical assessment of patterns of walking and muscle tone.

5.3.2 Experimental considerations.

i. The subjects lay comfortably prone on a couch watching a video of their own choosing in a warm quiet room in the presence of their parents.

ii. The feet were off the couch allowing a full knee-joint range from full extension to full flexion.

iii. The normal and affected limbs were compared.

iv. Surface electromyography (EMG) was assessed with the subject at rest. Disposable surface electrodes (Medicotest "Blue Sensor") with a diameter of 5 mm were arranged in a bipolar array with an inter-electrode distance of approximately 6-10 cm, depending on the size of the child, along the long axis of the muscle, with the active electrode proximal to the reference. The earth electrode was placed either on the same or opposite limb. The skin was cleansed with an abrasive cream prior to electrode placement. The quadriceps/ hamstrings muscles were studied as agonist-antagonist pairs. Electrodes were placed over the middle anterior and posterior thigh overlying each muscle. Electrode jelly to improve contact was used to supplement that provided on the disposable electrode. A piece of adhesive tape secured the position of the electrodes to the skin overlying the muscle of interest. The EMG signal was acquired and displayed on an 8-channel SLE 800 EEG machine with a bandwidth

of 0.5-150 Hz using a paper speed of 1.5 cm/second and display gain of 10μV/mm.

v. Joint angles were measured using a flexible twin axis Penny and Giles goniometer with a +/- 5V output reduced to +/- 500μV by a variable resistor prior to connection with the SLE recorder. Each goniometer trace was sandwiched between the EMG signals of the muscles to allow EMG interpretation in relation to displacement amplitude and angular velocity of ramp stretch or frequency of sinusoidal oscillation. The goniometer output was calibrated with a plastic goniometer which was used to give a calibration signal to the SLE recorder of 40° peak to peak and allowed cross-reference with the goniometer's own instantaneous liquid crystal display. The signal error was +/- 1-2°.

The quadriceps and hamstring muscles were tested over a 90° range, 0° representing full knee extension with the goniometer taped across the lateral aspect of the knee joint.

Figure 5.3.2.1 shows the principal joint positions (axes of rotation) of 15°, 45° and 75° about which the muscles were stretched over an approximately 30° arc.

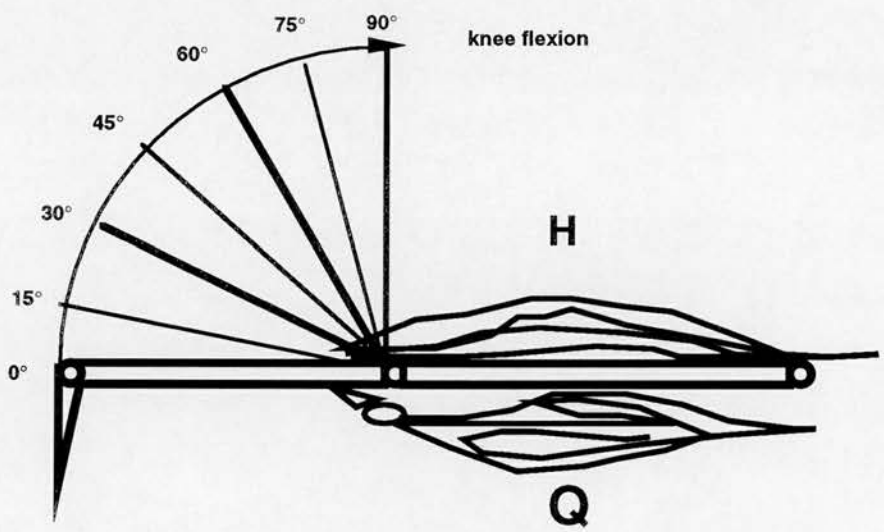


Figure 5.3.2.1 . Knee position and muscle length.
The resting lengths of quadriceps (Q) and hamstring (H) muscles depends on the position of the knee joint. As the knee moves from full extension at 0° to 90° of flexion the quadriceps are lengthened and the hamstrings shortened and vice versa in moving from 90° of flexion to full knee extension (0°).
Phasic ramp stretch : each muscle was rapidly stretched over about a 30°amplitude. This was from 0-30°, 30-60° and 60-90° of knee flexion during quadriceps stretch and 90-60°, 60-30° and 30-0° for rapid hamstring stretch. The reflex angular velocity threshold for quadriceps and hamstrings was measured in °/s.
Sinusoidal stretch : The knee joint was oscillated for aproximately 30° about a centre of oscillation represented by the dashed lines. Each centre of oscillation corresponded to 15°, 45° and 75° of knee flexion. The reflex frequency threshold for quadriceps and hamstrings was measured in Hz.

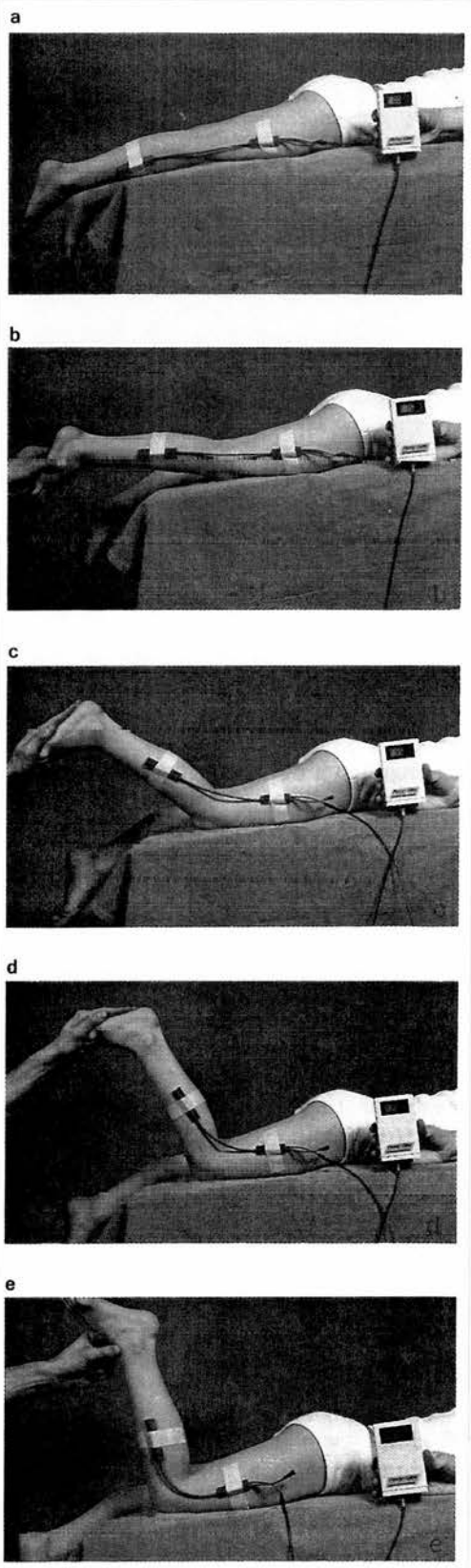


Figure 5.3.2.2 a-e.
Measurement of dynamic knee joint angles
The clinical attachment of the flexible goniometer across the knee joint to measure sagital-plane motion at the knee with the subject prone:
a. knee extended. **b.** flexed to 15°,
c. flexed to 45°, **d.** flexed to 75° **e.** flexed to 90°.
To avoid confusion the surface EMG electrodes are not included in these photographs. As the knee is flexed, the quadriceps lengthen and the hamstrings shorten and vice-versa.
Ramp stretches were performed from 0°-30°, 30°-60° and 60°-90° for quadriceps ramp stretches and in reverse direction from 90°-60°, 60°-30° and 30°-0° for hamstring ramp stretches.
Sinusoidal stretches of + / - 15° were imposed at centres of oscillation of 15°, 45°and 75° .
By permission.Lin, Brown and Brotherstone, 1994a.

5.3.3 Stretch protocols.

5.3.3.1. Ramp stretches.

Each muscle was subjected to sudden manual ramp stretches at amplitudes of about 30° displacement at varying manually-delivered angular velocities of stretch. For the quadriceps, the ramp flexions applied to quadriceps spanned approximately 0°-30°, 30°-60° and 60°-90°. For hamstrings, ramp extensions from 90°-60°, 60°-30° and 30°-0° were applied. For both muscle groups, ramp stretches began with the muscles at their shortest lengths with the hip at neutral, ie neither flexed nor extended.

5.3.3.2. Sinusoidal stretches.

Each muscle underwent manual sinusoidal stretch at rest according to the method of Burke *et al* 1971, at amplitudes of +/- 15° about three centres of oscillation: 15°, 45° and 75° of knee flexion.(figs.5.3.2.1 and 5.3.2.2 b, c, d). Reflex excitability is known to vary with initial muscle length (Burke *et al* 1971). The centre of oscillation determines the starting length of the muscle and allows comparison of reflexes length for length between subjects. For any pair of agonist / antagonist muscles, the shortest position of the one is the longest position of the other. The frequency of sinusoidal oscillation varied from 0.5-3.5Hz at the knee.

Conformity to a sinusoidal waveform was maintained according to the goniometer output trace. The torque required to displace the limbs at any given amplitude and frequency was applied manually by the examiner but was not measured. Since the accelerations required for an oscillating object varies with the square of the frequency (see section 9 and Walsh, 1992), only relatively low frequencies of stretch could be delivered with a manual stretch.

In this context, it should be remembered that the resonant frequency of the normal adult knee at 'moderate amplitudes of motion' is approximately 0.5Hz (Walsh, 1992, table 11.1, p181): the resonant or natural frequency corresponds to the frequency at which the amplitude of motion is maximal. For the purpose of these experiments, oscillations of up to seven times the normal adult natural frequency were manually delivered.

5.3.4 Wartenberg's Pendulum Test.

The pendular swing of the leg at the knee joint after eliciting the knee jerk has been examined by Burke, Gillies and Lance (1970) and reviewed by Burke and Lance (1973).

Knee jerks were elicited with the children dangling their legs over the edge of the

couch with goniometer and EMG monitoring. The maximum duration of the damped oscillation in seconds and maximum amplitude of swing in degrees following a tap to the patella tendon were measured. Traces which showed evidence of EMG activity at rest or 'restlessness' before or after taps were discarded. Pendulum studies were also performed by allowing the leg to drop from full knee extension with the subject seated. This allowed standardised stretch stimuli to the quadriceps muscles (assuming similar inertial characteristics between limbs). The combined studies took approximately 1 hour for the thigh muscles of both legs for each child.

5.3.5 EMG analysis.

All the tests were performed by the same examiner (JPL) and electrode placement and operation of the SLE recorder under the supervision of the same technician (RB). All the traces of EMG activity were analysed according to the following criteria:

1. In the case of the sinusoidal stretches, the phase relations of the EMG discharges relative to the angular displacement was examined. According to the methods described by Burke, Andrews and Gillies (1971), for velocity-dependent reflex EMG discharges evoked by sinusoidal stretches, the peak EMG occurs at 90° in phase advance of the peak angular displacement, ie when the angular displacement and acceleration are zero (fig. 5.3.51b). The same engineering principles dictate that the peak reflex perturbation produced by a reflex EMG discharge occurs with a 180° phase lag to the peak EMG. On the other hand, a peak reflex EMG occurring at the time of peak angular displacement indicates a length-dependent, tonic response (fig. 5.3.5.1c), commonly encountered when the muscle was voluntarily or involuntarily active at the start of stretch or during the stretch cycle .

Figure 5.3.51a-d attempts to illustrate a number of commonly encountered motor patterns: **a.** voluntary alternating movements, **b.** passive movements eliciting a velocity-dependent stretch reflex (ie phasic reflex), **c.** passive movements eliciting a length-dependent stretch reflex (ie tonic reflex)and finally **d.** passive motion imposed on a pre-activated or tonically contracting muscle. Irrespective of whether the EMG discharges are primary or secondary, ie, voluntary, involuntary or reflex, the EMG always occur 180° in phase advance of the generated motion pattern. The difficulty arises when the primary motion pattern elicits a reflex discharge (either phasic or tonic) which then alters the subsequent motion pattern: in this context, secondary or reflex EMG discharges may be detectable.

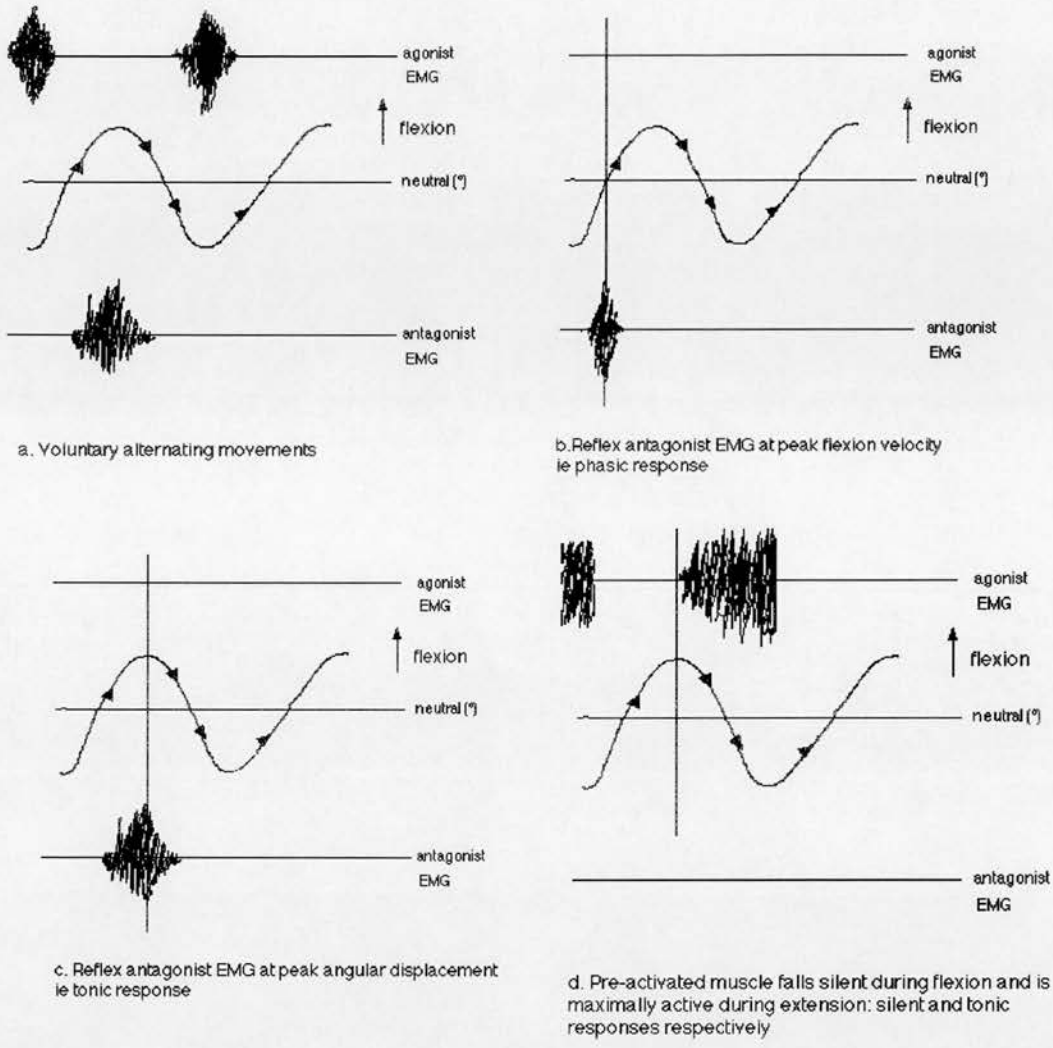


Figure 5.3.5.1 a-d Schematic representation of timing and effects of EMG activity

a. Voluntary alternating movements: the EMG bursts occur 180° *in phase advance* of angular displacement which is the same as saying that the angular displacement always has a 180° phase lag with the EMG discharge. The joint angle trace represents active movement

b. Velocity -dependent reflex EMG discharge of antagonist during passive flexion occurs at peak velocity (vertical line): a so-called *phasic response.*

c. Length-dependent reflex EMG discharge of antagonist occurs at peak angular displacement (vertical line): a *tonic response.*

d. The agonist muscle is active before passive flexion and becomes silent during flexion: so-called *silent period* coinciding with spindle unloading. The muscle resumes activity (after the vertical line) during extension and reaches a maximum at peak angular extension: a*tonic response.*

In the case of fig. 5.3.5.1**b**, the evoked velocity-dependent reflex would cause a distortion of the angular displacement with a 180° phase lag (ie downstream) of the actual discharge itself and this is sketched in as a steeper extension motion. In the case of fig. 5.3.5.1**c**, the distortion to the passive motion also occurs with a 180° phase lag with respect to the EMG discharge and this too causes excessive extension and delays flexion. One result of the situation in fig. 5.3.5.1d, is that the unopposed flexion component of motion would be longer than the opposed extension component. An obvious common clinical example would be a combination of figs 5.3.5.1a and b, in which the primary motion generates a stretch reflex which then disturbs subsequent motion, for instance, upsetting the usual pre-positioning of the foot in terminal swing to produce a toe-contact pattern instead of the usual heel-strike. Such an equinus foot strike would then cause an initial passive dorsiflexion which could result in a reflex plantarflexion causing instability in the single-limb stance phase of the gait and so on. The final motion pattern being quite chaotic in relation to the expected pattern.

2. EMG silence was a pre-requisite prior to either ramp or sinusoidal stretching: sequences containing EMG activity prior to stretch were discarded as representing inadequate relaxation or involuntary activity (figs 5.3.5.2B, 5.3.5.3a, b).

3. Voluntary or involuntary co-contractions during sinusoidal stretch abolish or distort displacement and show augmented tonic EMG at maximum stretch and an EMG silent period (figs. 5.3.5 2A and 5.3.5 3a, b) when the muscle is unloaded (Angel 1973; Struppler, Burg and Erbel,1973). There is no way of knowing if the EMG represents a dystonic response (fig 5.3.5 3a) or a voluntary or involuntary EMG discharge (fig 5.3.5 3b)

4. During sinusoidal stretch, a triphasic agonist/antagonist EMG response was interpreted as characteristic of voluntary alternating flexion and extension movements This usually occurred at low frequency stretch cycles when subjects could anticipate the direction of movement and assisted the motion. Traces containing such activity were discarded.

5. Reflex excitability was measured in terms of the velocity (for ramp stretches)or frequency (for sinusoidal stretches) *threshold* at which the reflex EMG activity was excited. The reflex gain was established by the slope of the line obtained by plotting the reflex EMG against velocity during ramp stretches or frequency during sinusoidal stretches.

6. The initial muscle length was defined by the centre of oscillation about which a reflex

was elicited.The term "nonparetic" muscles refers to the unaffected limb of a hemiparetic child.

7. Nonparetic and hemiparetic reflex thresholds were compared at equivalent initial joint positions

5.3.6 Analyses.

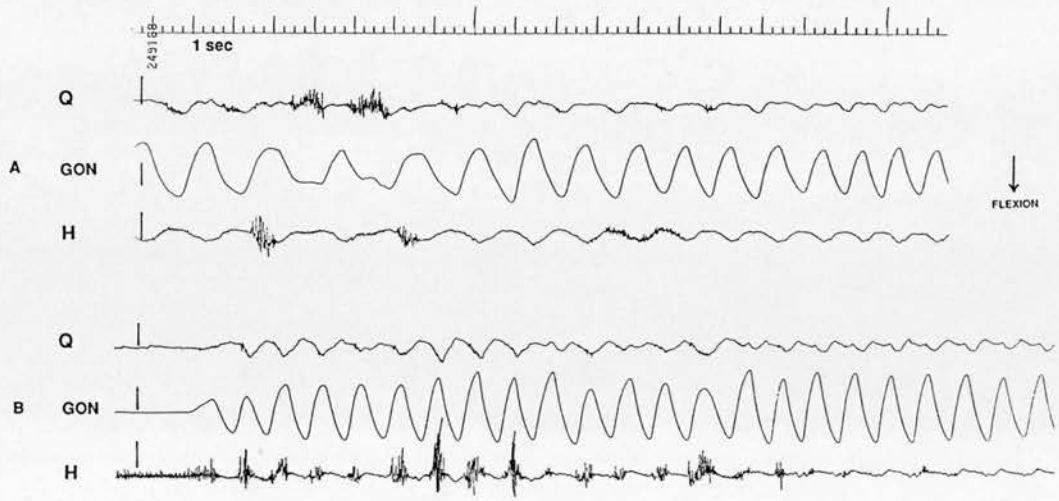
Qualitative analysis by examination of polygraphic traces to ensure comparability between limbs and to exclude 'contaminated' traces: ie traces which showed evidence of background EMG activity prior to the onset of stretch or during stretches.

The mean reflex EMG thresholds in degrees/second (°/s) for ramp stretches between 0°-30°, 30°-60° and 60°-90°: ie with the initial quadriceps muscle length short, intermediate or long. Sinusoidal stretches allowed the reflex frequency threshold to be measured in Herz (Hz) at equivalent centres of oscillation of 15°, 45° and 75°. For each stretch protocol, velocity and frequency thresholds were computed for each muscle and at each muscle length or centre of oscillation and compared using 95% confidence intervals and a paired **t** test (Statview+ Graphics). A similar comparison was made for limbs for which no reflex threshold could be reached. Results are given in tabulated form and graphically.

5.4 Results of ramp stretches.

5.4.1 Qualitative considerations

Quadriceps and hamstring muscles were subjected to rapid ramp stretch from rest in twelve cases. Results are expressed in terms of the minimum angular velocity threshold required to elicit a reflex EMG response. Figure 5.4.1 shows the EMG and goniometer traces from a 10 year old girl with a congenital right hemiplegia for nonparetic and hemiparetic quadriceps ramp stretches at three successive knee joint positions.



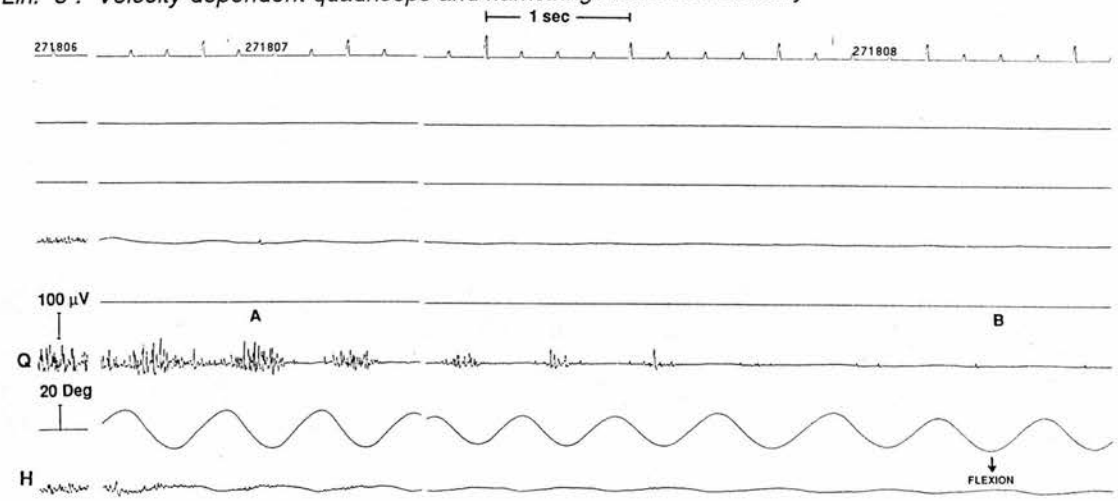
Passive sinusoidal flexion / extension of the Right knee joint at 0.5 - 1 Hz, 92 - 151°/sec

Case 18. Congenital Left Hemiplegia. Age 12.
Centre of oscillation: 15° of flexion from full extension.

A. Intermittent reflex EMG of Hamstrings and Quadriceps: normal response.
B. Stretch imposed on contracting Hamstrings demonstrates normal reflex response.

Q: Quadriceps EMG. H: Hamstrings EMG. GON: Goniometer.
Calibration bars: EMG 100μV, goniometer 20°, time 1 sec.

Figure 5.3.5.2 Nonparetic and hemiparetic EMG with passive sinusoidal stretch.
A. Effect of isolated active muscle contractions during passive manual sinusoidal stretches: the discharges alter the motion pattern by reducing peak passive extension during hamstrings (H) discharges and peak flexion during quadriceps (Q) discharges. Without background muscle activation, the passive motion does not elicit a reflex response. Silent muscle is relatively inexcitable.
B. effect of sinusoidal stretching imposed on background muscle activation: the hamstrings (H) are active prior to the onset of stretch, fall silent during flexion and the EMG pattern is augmented during extension. The hamstrings EMG amplitude appears to be proportional to the amplitude of passive motion, indicating a tonic response. Note that at the end of the trace, only a brief phasic hamstrings EMG discharge occurs in the absence of background hamstrings activity.



271808

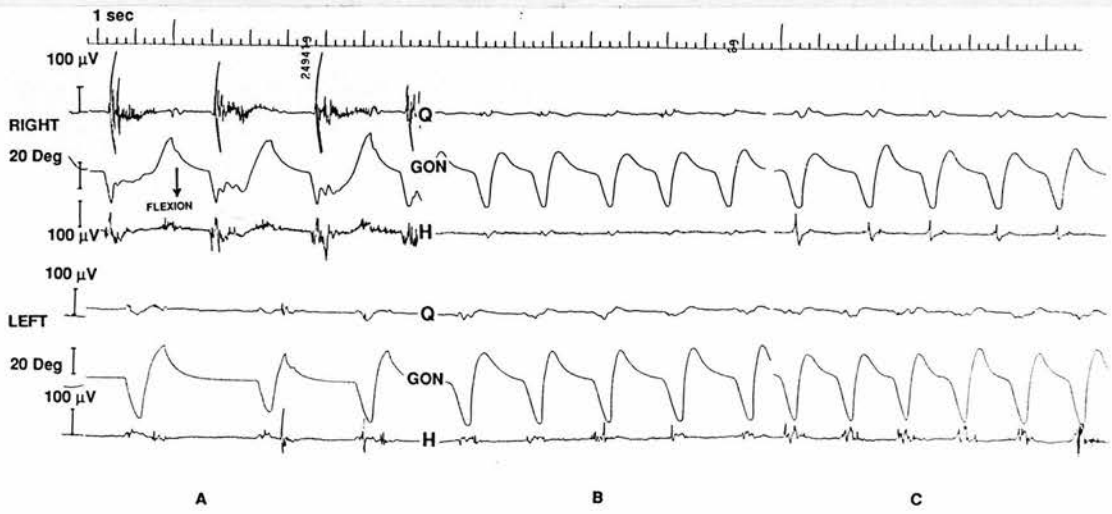
Congenital left hemidystonia in 12 year old girl.

Alternating passive sinusoidal left knee flexion / extension at 1.4 - 2.0 Hz. Patient supine.

A. Incomplete relaxation and appearance of tonic spasticity. B. Fully relaxed with electrical silence.

Q: quadriceps, H: hamstrings, GON: goniometer. Paper speed 6 cm/sec.

Figure 5.3.5.3 Passive muscle stretches in a case of left hemidystonia
Dystonic limb. Loading and unloading (silent) responses of the quadriceps (Q) and hamstrings (H) muscles in a dystonic limb when the muscle is active prior to stretch (A). The same passive sinusoidal stretches evoke no EMG response when there is no background activity (B).Note the expanded time-base. The trace has been cut and represents a join between two adjacent pages of recording.



249428/419/423/425/472/473/475

Congenital Right Hemiplegia in 10 year old girl.

Comparison of rapid knee flexion of right and left legs.

Starting angle.	Speed of flexion Deg/sec.	
	R	L
A. 0-30	143	124
B. 30-60	115	112
C. 60-90	102	118

No appreciable quadriceps phasic response of the normal limb.

Q: quadriceps, H: Hamstrings, Gon: goniometer. Paper speed 1.5 cm/sec

Figure 5.4.1 Reflex velocity threshold following phasic ramp stretch.

5.4.2 Quadriceps reflex velocity threshold with muscle length.

Table 5.4.a gives the results of the mean and standard deviation (SD), 95% confidence intervals (95%CI) and paired **t** test values of reflex velocity thresholds for the nonparetic and hemiparetic quadriceps muscles of the children according to initial muscle length. This reduced velocity sensitivity with increasing muscle length achieved statistical significance for the hemiparetic quadriceps ($p < 0.01$) but the nonparetic quadriceps in whom a velocity threshold could be elicited, did not show a significant length-dependent inhibition. Even though the graphical plots of mean reflex velocity thresholds (depicted without SD ranges) suggest that the nonparetic quadriceps are length sensitive, this did not achieve statistical significance. The nonparetic quadriceps reflex was refractory to ramp stretches from 0°-30° in 4/12 cases; from 30°-60° in 7/12 case and from 60°-90° in 9/12 cases indicating an increase in the reflex velocity threshold with increasing quadriceps length.

No differences in reflex velocity thresholds between nonparetic and hemiparetic quadriceps muscles could be established at comparable knee joint positions (Table 5.4.b, see also figs 5.4.2a and 5.4.3a).

Table 5.4.a. Comparison of the mean reflex velocity thresholds for the normal and hemiparetic quadriceps (Q) and hamstring (H) muscles during rapid (phasic) ramp stretch at varying knee-joint angles (muscle length) in childhood hemiplegia.

Muscle		N° cases (n=13)	Ramp stretch (°) KF	Reflex threshold reached	Velocity ramp stretch (°/s) mean	SD	Confidence int. Lower 95%	Upper 95%	Paired t-test p value
Normal	Q	8	0-30°	+	131.3	33.72	103.1	159.5	
Normal	Q	5	30-60°	+	146	31.29	107.1	184.9	NS
Normal	Q	2	60-90°	+	208.5	61.51	-344.2	761.2	*
Hemi	Q	10	0-30°	+	124.7	28.12	104.5	144.8	
Hemi	Q	6	30-60°	+	148.3	34.79	111.8	184.8	$p < 0.01$
Hemi	Q	5	60-90°	+	136.6	33.99	94.3	178.8	NS
Normal	H	8	30-0°	+	112.8	22.1	94.3	131.4	
Normal	H	7	60-30°	+	131.5	37.1	97.1	165.9	$p < 0.01$
Normal	H	4	90-60°	+	119	40.24	54.96	183	NS
Hemi	H	8	30-0°	+	101.1	22.5	82.2	119.9	
Hemi	H	7	60-30°	+	130	36.34	96.3	163.6	$p < 0.01$
Hemi	H	7	90-60°	+	156.7	55.6	105.2	208.2	$p < 0.02$

* = insufficient numbers for paired t test.
KF= knee flexion.

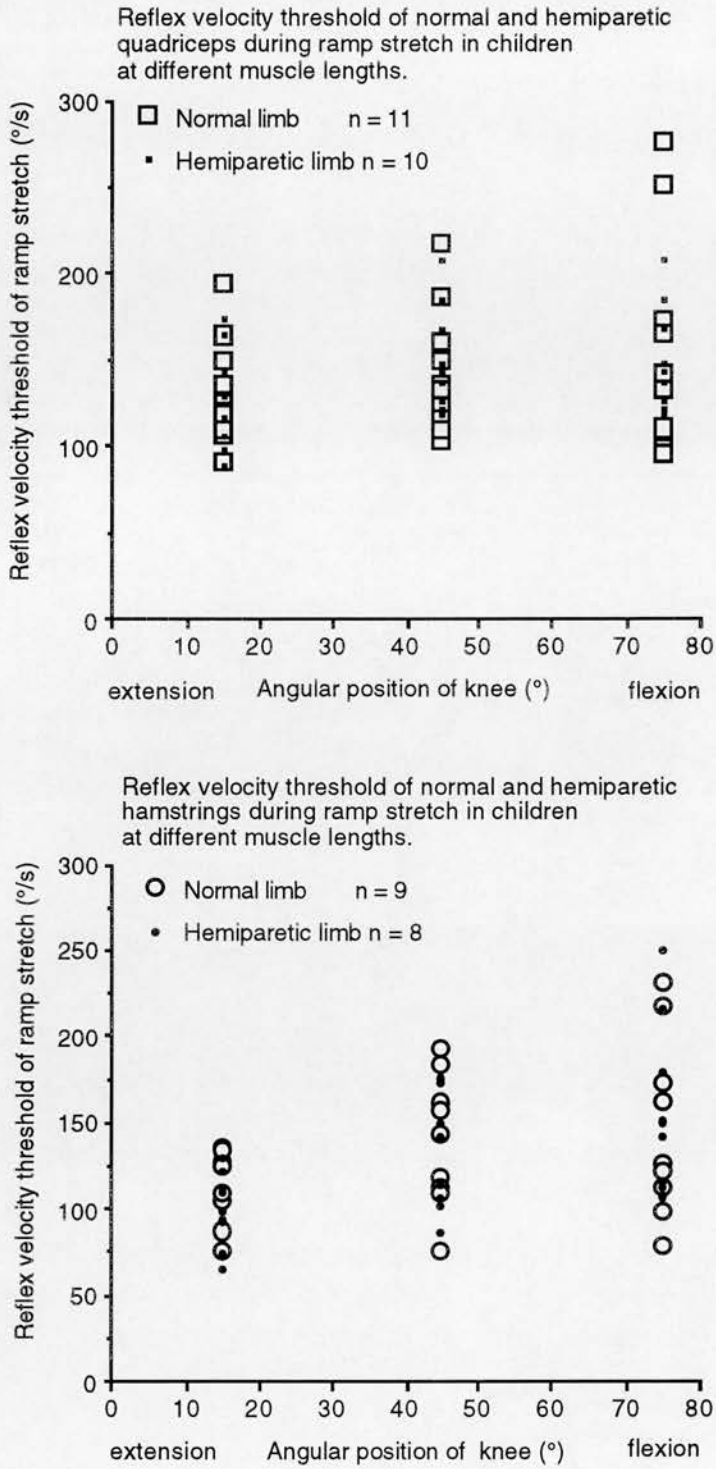


Figure 5.4 2 a-b. Reflex velocity thresholds and the joint angle.
Above:**a.** Nonparetic and hemiparetic quadriceps muscles and.
Below: **b.** Nonparetic and hemiparetic hamstrings muscles.

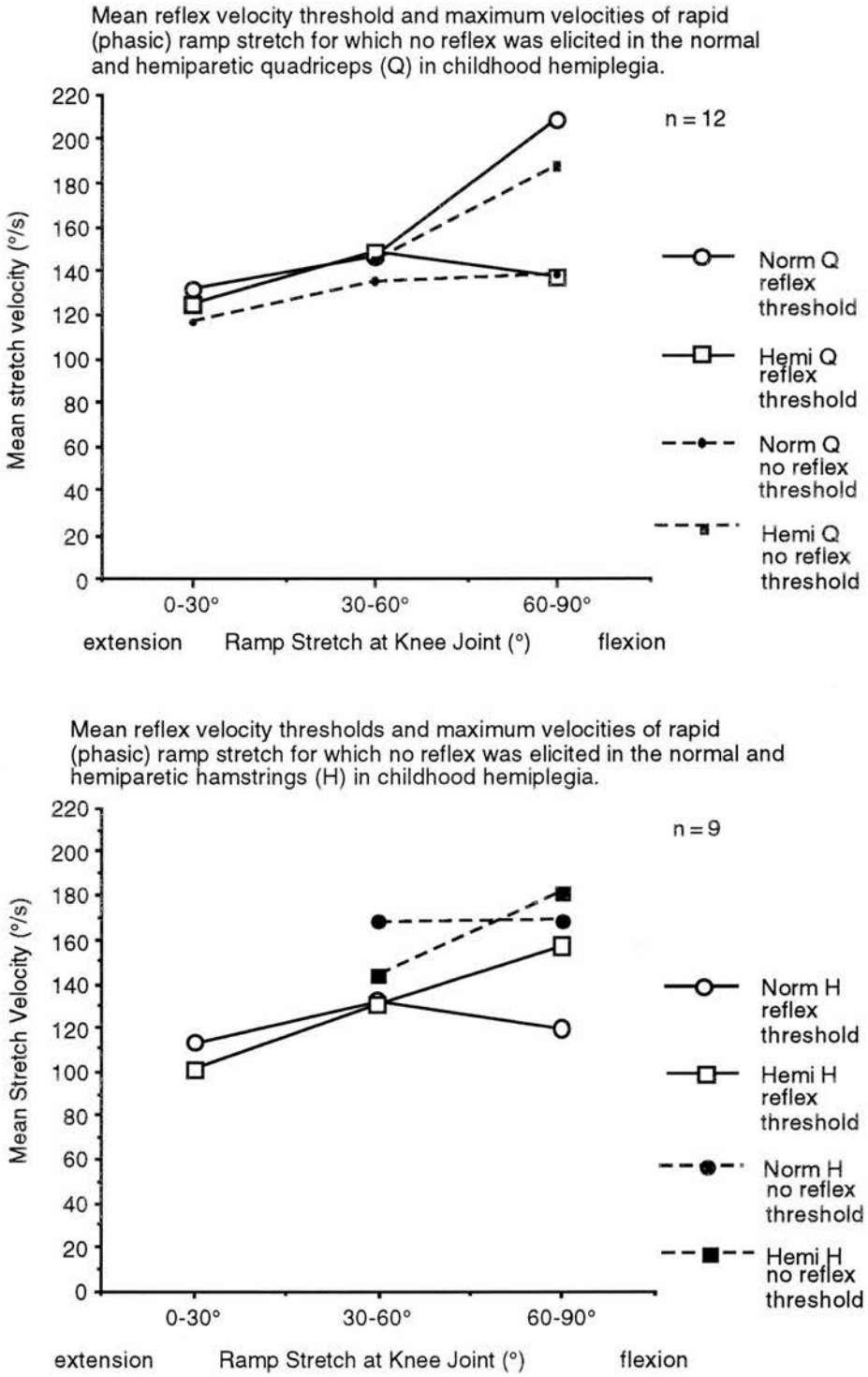


Figure 5.4.3a-b. Comparison of excitable and inexcitable thigh muscles.
Threshold velocities and velocities of maximum stretch in **a.**quadriceps (above) and **b.** hamstrings (below)

Table 5.4.b. Mean reflex velocity thresholds and maximum velocities for which no reflex threshold was reached following rapid (phasic) ramp stretch of quadriceps (Q) and hamstrings (H) muscles in childhood hemiplegia.

Muscle		N° cases	Ramp stretch (°) KF	Reflex threshold reached	Velocity ramp stretch (°/s) mean	SD	Confidence int.. Lower 95%	Upper 95%	Paired t-test p value
Normal	Q	8	0-30°	+	131.3	33.72	103.1	159.5	
Hemi	Q	10	0-30°	+	124.7	28.12	104.5	144.8	NS
Normal	Q	4	0-30°	-	116.7	25.34	76.42	157.0	
Hemi	Q	0	0-30°	-	----	----	----	----	*
Normal	Q	5	30-60°	+	146.0	31.29	107.1	184.9	
Hemi	Q	6	30-60°	+	148.3	34.79	111.8	184.8	NS
Normal	Q	7	30-60°	-	135	37.72	100.1	169.8	
Hemi	Q	4	30-60°	-	144.4	28.89	98.5	190.4	NS
Normal	Q	2	60-90°	+	208.5	61.51	-344.2	761.2	
Hemi	Q	5	60-90°	+	136.6	33.99	94.3	178.8	*
Normal	Q	9	60-90°	-	138.2	57.16	94.2	182.1	
Hemi	Q	5	60-90°	-	187.2	73.18	96.3	278.0	NS
Normal	H	8	30-0°	+	112.8	22.1	94.3	131.4	
Hemi	H	8	30-0°	+	101.1	22.5	82.2	119.9	NS
Normal	H	0	30-0°	-	----	----	----	----	
Hemi	H	0	30-0°	-	----	----	----	----	*
Normal	H	7	60-30°	+	131.5	37.1	97.1	165.9	
Hemi	H	7	60-30°	+	130.0	36.34	96.3	163.6	NS
Normal	H	2	60-30°	-	167.5	34.64	-143.8	478.8	
Hemi	H	1	60-30	-	143	----	----	----	*
Normal	H	4	90-60°	+	119.0	40.24	54.96	183.0	
Hemi	H	7	90-60°	+	156.7	55.6	105.2	208.2	NS
Normal	H	5	90-60°	-	168.8	53.93	101.8	235.7	
Hemi	H	1	90-60°	-	180	----	----	----	*

\+ = Reflex velocity threshold reached. - = No reflex velocity threshold reached.
* = Insufficient numbers for paired t test. KF= knee flexion

5.4.3 Hamstring reflex velocity threshold with muscle length.

The hamstring reflex velocity threshold diminishes with increasing muscle length, i.e as the knee position moves towards full extension, the hamstrings become more excitable, and this is moderately significant for the nonparetic ($p < 0.01$) and hemiparetic ($P < 0.01$) hamstrings (Table 5.4.a). Nonparetic and hemiparetic hamstrings appeared to have a similar reflex velocity threshold at comparable muscle lengths (Table 5.4.b, figs. 5.4.2b and 5.4.3b).

5.4.4 Variations in reflex velocity threshold with initial muscle length.

Table 5.4.b indicates those nonparetic and hemiparetic quadriceps and hamstring muscles for which no reflex velocity threshold could be elicited. At each range of stretch, these muscles were subjected to either similar or increased velocities of stretch when compared with muscles for which a reflex response was obtained. The proportion of muscles for which it was not possible to elicit a reflex velocity threshold increased with increasing quadriceps length and hamstring shortening respectively. Overall, the quadriceps and hamstring muscles were most velocity-sensitive, close to the position of full knee extension but nonparetic and hemiparetic muscle groups had similar reflex velocity thresholds (Table 5.4.b, figs. 5.4.1, 5.4.2 and 5.4.3).

5.5 Results of sinusoidal stretching.

5.5.1 Qualitative examination of EMG discharges during sinusoidal stretches.

Sinusoidal stretching of quadriceps and hamstring muscles was performed in thirteen children. Results are expressed as the minimum frequency of sinusoidal oscillation required to elicit a reflex EMG response in the quadriceps and hamstrings: the *reflex frequency threshold*. The centre of oscillation determines the initial muscle length (figures 5.3.2.1 and 5.3.2.2 a-e). The amplitude of manual sinusoidal displacement about any given centre of oscillation was 20°-50°, keeping as close to 30° as possible.

Figure 5.5.1a shows the EMG and goniometer output during passive sinusoidal stretches at varying frequencies and muscle lengths of the nonparetic quadriceps and hamstrings of a 12 year old boy with a congenital left hemiparesis, while figure 5.5.1b shows the effects of sinusoidal stretching of the hemiparetic limb in the same child. The nonparetic quadriceps and hamstrings appear inexcitable at all centres of oscillation and frequencies of sinusoidal stretch which were varied from 0.5 to 2Hz (fig. 5.5.1a). Close to full knee extension (centre of oscillation of 15°) and at sinusoidal stretch frequencies of 0.5-0.75Hz (ie at approximately 80°/s), the hemiparetic quadriceps appeared to demonstrate low amplitude tonic stretch reflex in response to flexion which deformed the terminal flexion goniometer trace and assisted extension of the knee. The first phasic quadriceps response occurring at 1Hz (90-120°/s) and the amplitude of this phasic response increasing with the rising frequency of passive stretches applied. This demonstrates a frequency gain. At the same centre of oscillation, close to full knee extension, there is a brief hamstrings tonic discharge at

0.5-1Hz of sinusoidal stretch, the hamstrings only responding with a phasic discharge at frequencies of 1.3-2Hz, again showing a reflex frequency gain. As the knee joint angle was flexed from 45° through to 75°, so the tonic stretch reflex of quadriceps muscle, which appears as modulation of the background EMG activity, increases in amplitude. The hamstrings phasic stretch reflex appears to decrease with increasing knee flexion since it can only be elicited at the maximum frequencies of 1.5-2Hz about 75° of knee flexion.

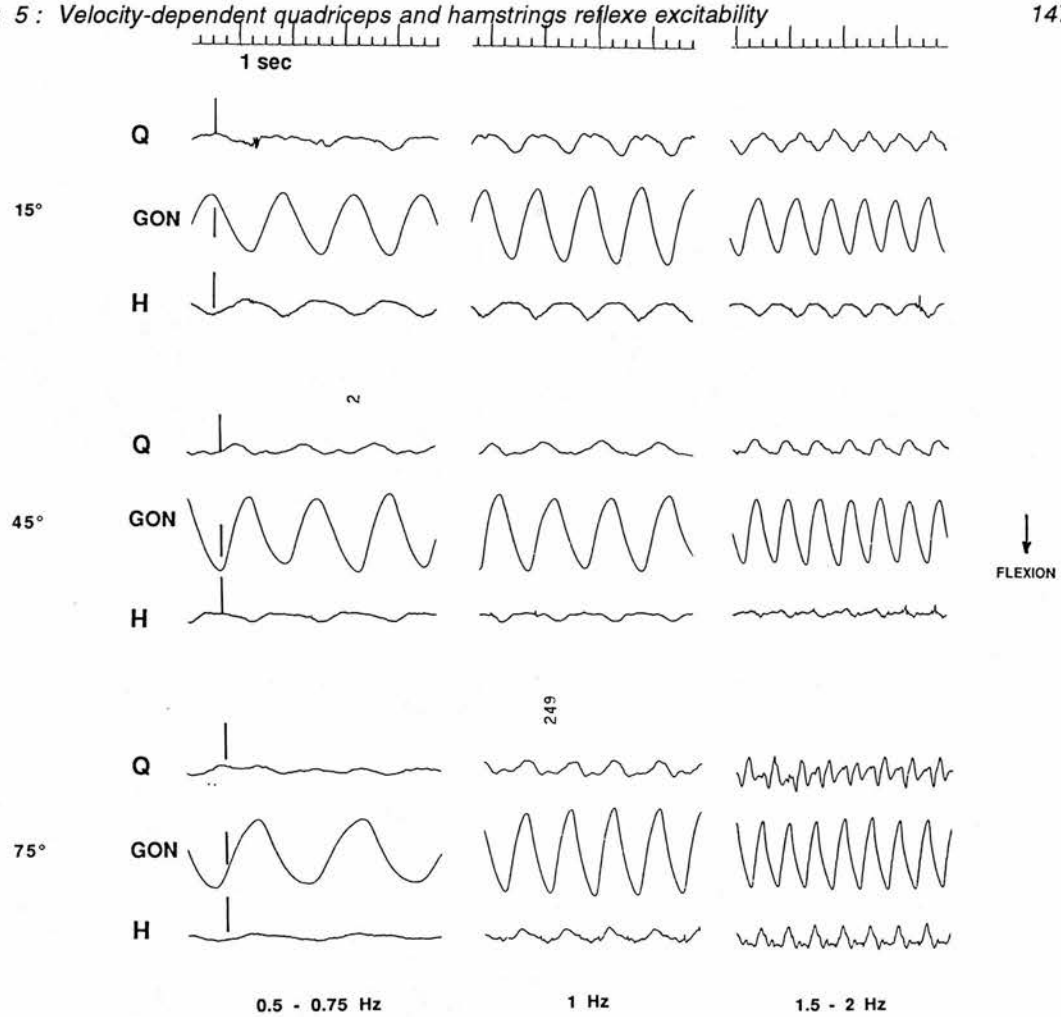
5.5.2 Quadriceps reflex frequency threshold and muscle length.

The mean reflex frequency threshold for nonparetic and hemiparetic quadriceps muscles in 13 children is shown in table 5.5.a at three knee joint positions (centres of oscillation) of 15°, 45° and 75° of knee flexion respectively. As for ramp stretches, the number of cases in whom a reflex frequency threshold could be established declined as the centre of oscillation approached 90° of flexion. There were no differences in reflex frequency threshold between nonparetic and hemiparetic quadriceps at any given muscle length (figure 5.5.2 and table 5.5.b).

5.5.3 Hamstrings reflex frequency threshold and muscle length.

Similar mean sinusoidal stretch frequencies elicited a hamstring reflex at the three centres of oscillation (table 5.5a). There was a weakly significant lowering of the hamstrings reflex frequency threshold on paired **t** testing for hemiparetic compared with nonparetic hamstrings ($p < 0.05$) close to full knee extension (table 5.5.b), but this difference is not seen at shorter hamstring positions.

For sinusoidal oscillation, as with phasic ramp stretching, the number of cases in which it was possible to establish a reflex frequency threshold in quadriceps and hamstrings declined with increasing knee flexion. The maximum frequencies of sinusoidal stretch to which these muscles were exposed were similar to those in which a threshold response was obtained (table 5.5.2).



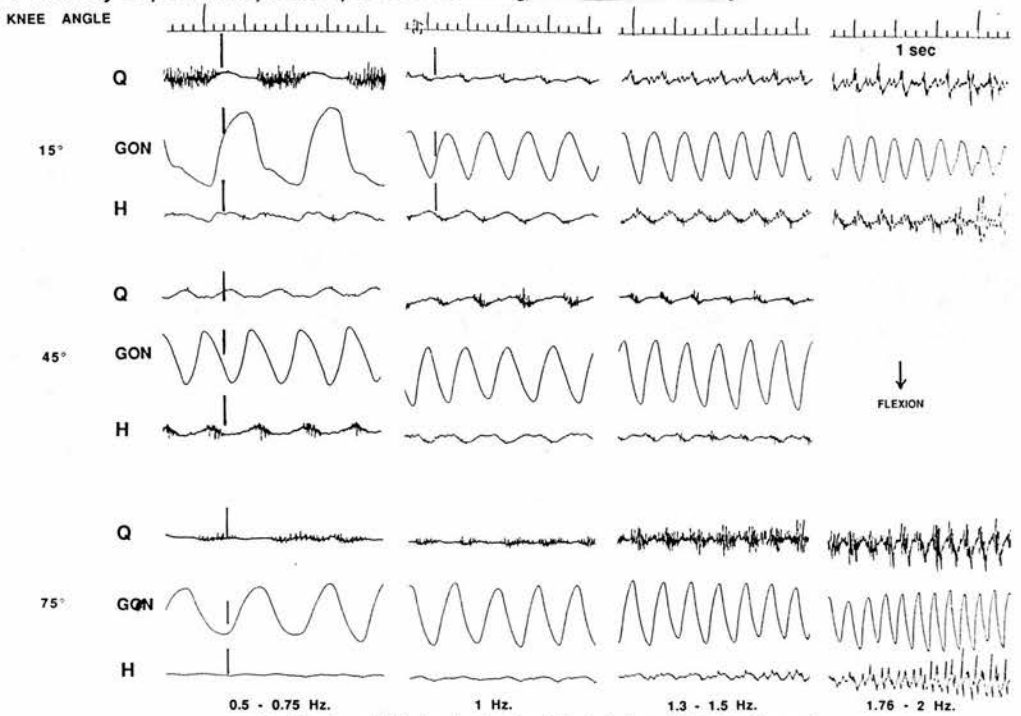
Passive sinusoidal flexion / extension of the Right knee joint at varying angles.

Case 18. Congenital Left Hemiplegia. Age 12.
Centres of oscillation: 15°, 45° and 75° of flexion from full extension.
Displacement amplitudes 30-50°.
Velocities: 50-90°/s at 0.5-0.75 Hz; 90-120°/s at 1 Hz; 100-168°/s at 1.5-2 Hz.

EMG in Quadriceps and Hamstrings silent for all angles and frequencies.

Q: Quadriceps EMG. H: Hamstrings EMG. GON: Goniometer.
Calibration bars: EMG 100μV, goniometer 20°, time 1 sec.

Figure 5.5.1a Reflex discharges of nonparetic limb with sinusoidal stretch.



Passive sinusoidal flexion / extension of the Left knee joint at varying angles.

Case 18. Congenital Left Hemiplegia. Age 12.
Centres of oscillation:15°, 45° and 75° of flexion from full extension.
Displacement amplitudes 30-60°. Velocities: 80°/s at 0.5-0.75 Hz; 70 - 95°/s at 1 Hz; 90 - 120°/s at 1.3 - 1.5 Hz; 135°/s at 1.76 Hz and 235°/s at 2Hz.

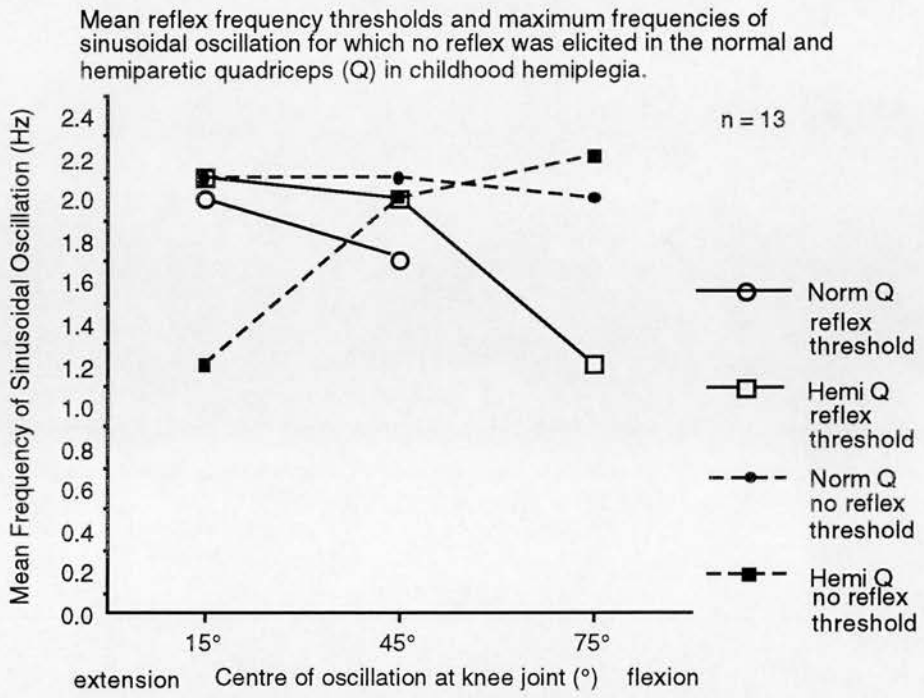
EMG in Quadriceps EMG most active at *shortest* postion i.e about 15° knee flexion. Hamstrings EMG most active at *longest* position i.e 15° knee flexion. Note quadriceps EMG in phase with maximum stretch at slow velocities but moving *in phase advance* of stretch at higher velocities.
Q: Quadriceps EMG. H: Hamstrings EMG. GON: Goniometer.
Calibration bars: EMG 100μV, goniometer 20°, time 1 sec.

Figure 5.5.1b Reflex discharges of hemiparetic limb with sinusoidal stretch.

Table 5.5.a Comparison of the mean reflex frequency thresholds for the normal and hemiparetic quadriceps (Q) and hamstring (H) muscles during sinusoidal oscillation at increasing muscle lengths in childhood hemiplegia.

Muscle	N° cases	Centre of oscillation (°) KF	Reflex thresh.	Frequency of oscillation (Hz) mean	SD	Confidence int.. Lower 95%	Upper 95%	Paired t-test p value
Normal Q	8	15°	+	2.0	0.59	1.5	2.5	
Normal Q	6	45°	+	1.7	0.37	1.3	2.1	NS
Normal Q	0	75°	+	----	----	----	-----	*
Hemi Q	6	15°	+	2.1	0.39	1.7	2.5	
Hemi Q	8	45°	+	2.0	0.62	1.49	2.5	NS
Hemi Q	1	75°	+	1.2	----	----	-----	*
Normal H	8	15°	+	1.8	0.54	1.3	2.2	
Normal H	6	45°	+	1.7	0.37	1.3	2.1	NS
Normal H	3	75°	+	2.0	0.83	0.0	4.1	NS
Hemi H	9	15°	+	1.3	0.34	1.1	1.6	
Hemi H	8	45°	+	2.0	0.6	1.4	2.5	p <0.05
Hemi H	5	75°	+	1.8	0.72	0.9	2.7	NS

* = insufficient numbers for paired t test. KF= knee flexion.



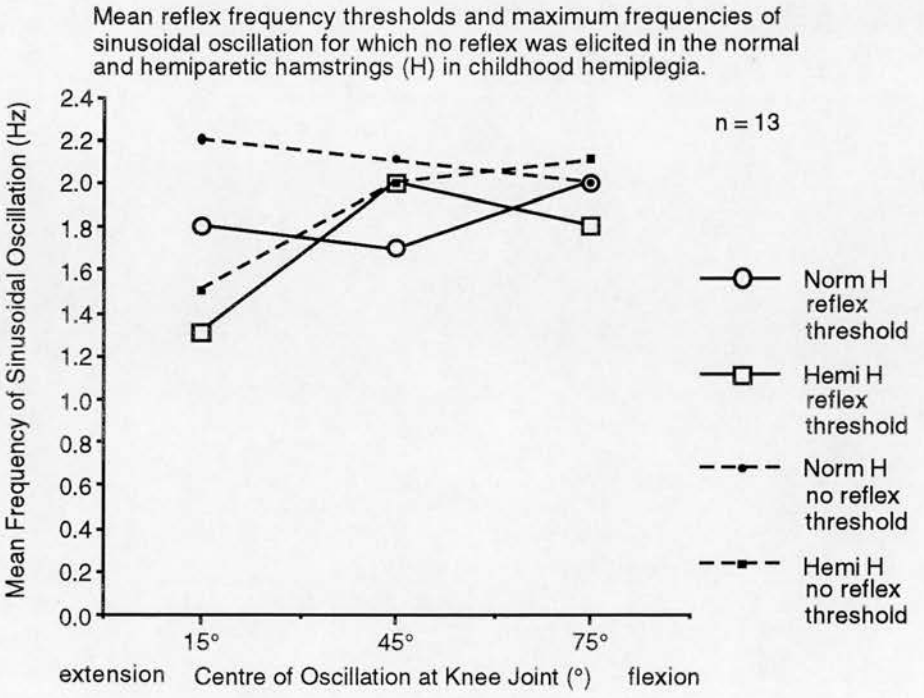


Figure 5.5.2 Relationship between joint angle and reflex frequency threshold.
Quadriceps muscles (top) and Hamstrings muscles (bottom). No real pattern emerges for the quadriceps muscles. The reflex frequency threshold appears to rise with decreasing muscle length for the hamstrings: ie they are most excitable close to full knee extension.

Table 5.5.b. Mean reflex frequency thresholds and maximum frequencies of sinusoidal oscillation for which no reflex was elicited in normal and hemiparetic quadriceps (Q) and hamstrings (H) in childhood hemiplegia.

Muscle		N° cases	Centre of oscillation reached mean (°) KF	Reflex thresh. SD	Frequency of oscillation (Hz) 95%	95%	Confidence int.. Lower p value	Upper	Paired t-test
Normal	Q	8	15°	+	2.0	0.59	1.5	2.5	
Hemi	Q	6	15°	+	2.1	0.39	1.7	2.5	NS
Normal	Q	5	15°	-	2.1	0.75	1.1	3.0	
Hemi	Q	7	15°	-	1.2	0.29	0.9	1.5	NS
Normal	Q	6	45°	+	1.7	0.37	1.3	2.1	
Hemi	Q	8	45°	+	2.0	0.62	1.49	2.5	NS
Normal	Q	7	45°	-	2.1	0.79	1.5	2.8	
Hemi	Q	4	45°	-	2.0	0.05	1.9	2.1	NS
Normal	Q	0	75°	+	----	----	----	----	
Hemi	Q	1	75°	+	1.2	----	----	----	*
Normal	Q	13	75°	-	2.0	0.44	1.7	2.3	
Hemi	Q	11	75°	-	2.2	0.55	1.8	2.5	NS
Normal	H	8	15°	+	1.8	0.54	1.3	2.2	
Hemi	H	9	15°	+	1.3	0.34	1.1	1.6	< 0.05
Normal	H	5	15°	-	2.2	0.97	1.0	3.4	
Hemi	H	4	15°	-	1.5	0.25	1.1	1.9	NS
Normal	H	6	45°	+	1.7	0.37	1.3	2.1	
Hemi	H	8	45°	+	2.0	0.6	1.4	2.5	NS
Normal	H	7	45°	-	2.1	0.72	1.5	2.8	
Hemi	H	4	45°	-	2.0	0.05	1.9	2.1	NS
Normal	H	3	75°	+	2.0	0.83	0.0	4.1	
Hemi	H	5	75°	+	1.8	0.72	0.9	2.7	NS
Normal	H	10	75°	-	2.0	0.35	1.7	2.3	
Hemi	H	7	75°	-	2.1	0.28	1.8	2.4	NS

+ = Reflex frequency threshold reached. - = No reflex frequency threshold reached.
* = Insufficient numbers for paired t test. KF= knee flexion.

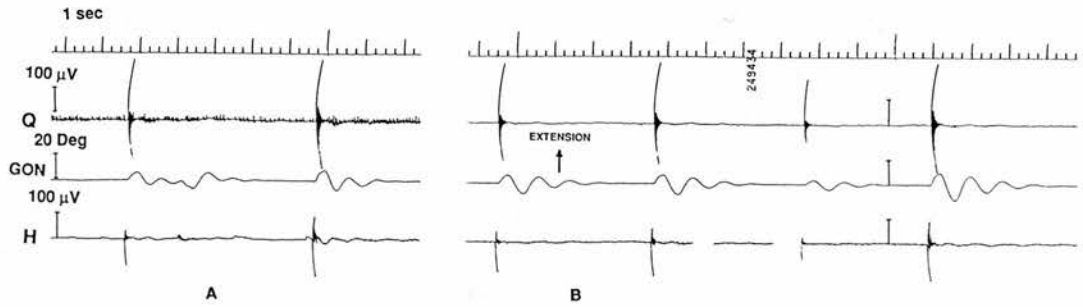
5.6 Comparison of ramp versus sinusoidal stretching.

Although rapid ramp stretches showed a close relationship between reflex velocity threshold and initial muscle length, no differences in reflex velocity threshold between nonparetic and hemiparetic limbs were apparent at any given muscle length. Sinusoidal stretching demonstrated a weakly significant lowering of the hamstring reflex frequency threshold in the hemiparetic compared with nonparetic hamstring at a position close to full knee extension only, but no such difference could be found for the quadriceps. Overall, the dominant feature was for the quadriceps and hamstrings muscles to be most excitable and have the lowest reflex thresholds close to the position of full knee extension, irrespective of whether the limb was paretic or nonparetic.

5.7 Wartenberg's Pendulum Test.

The free oscillation of the leg after a tendon tap to elicit the quadriceps reflex may give an idea of the excitability of the reflex arc and the stiffness of the subsequent muscular damping . The amplitude of the EMG discharge and initial knee extension would correspond to the magnitude of the reflex elicited, and the duration of the damped oscillations would represent an index of the muscle stiffness. Accordingly, knee jerks were elicited in 12 cases, excluding the case of hemidystonia, and the means and 99% confidence intervals for the duration of damped oscillation and maximum amplitude of swing are shown in table 5.7. There were no significant differences between sides. In 5 cases, one or other limb was restless during the test (figure 5.7.1) so that only 7/12 paired comparisons between nonparetic and hemiparetic legs were possible.

Figure 5.7.2 illustrates that a tendon jerk (A) is a more powerful phasic stretch reflex stimulus than the quadriceps stretch elicited by allowing the leg to fall from full extension (B) irrespective of limb status. The oscillation is of a greater amplitude and lasts almost twice as long on the nonparetic side (lower traces), irrespective of whether it is generated by a tendon jerk or a leg drop: the nonparetic thigh muscles are underdamped with respect to the hemiparetic limb. This indicates that in certain cases, such as above, the hemiparetic muscle exhibits an increase in the intrinsic viscoelastic stiffness, such a stiffness tending to dampen imposed motions (see section 4, Plastic properties of muscle).



249432/434

Congenital Right Hemiplegia in 10 year old girl.

Right knee jerk with free swing, subject seated.

A. Distorted by voluntary quadriceps EMG activity. B. Relaxed: damped oscillation lasting 3.25 seconds with maximum amplitude of 22 degrees.

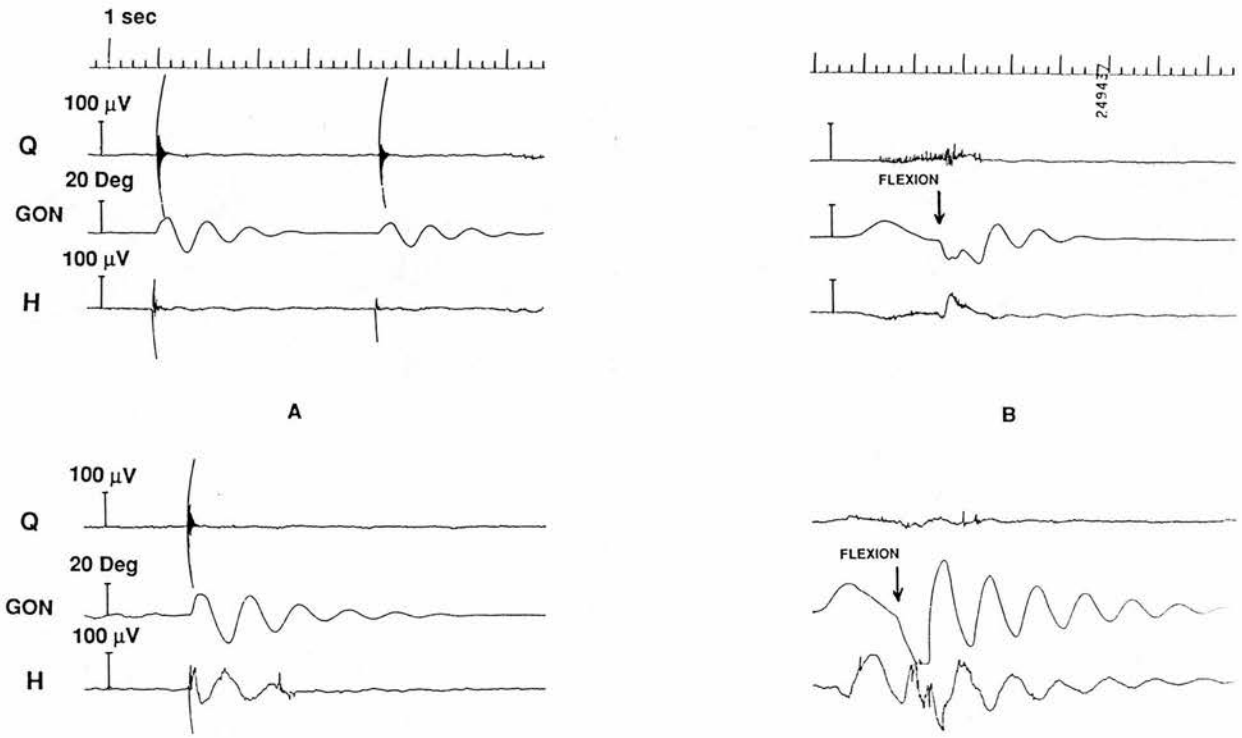
Q: quadriceps, H: Hamstrings, Gon: goniometer. Paper speed 1.5 cm/sec

Figure 5.7.1 Wartenberg's pendulum test following knee jerks.

Table 5.7.1. Wartenberg's pendulum test at the knee

Knee Jerk	NP n=10	HP n=9	Paired t Test n=7
Duration of Damped Oscillations (S)			
Mean	4.22	3.922	NS
SD	1.355	1.144	
99% Lower CI	3.11	2.94	
99% Upper CI	5.324	4.905	
Amplitude of Maximum Swing (°)			
Mean	21.62	17.8	NS
SD	16.77	8.72	
99% Lower CI	7.95	10.36	
99% Upper CI	35.28	25.34	

Comparison of duration of damped oscillation and maximum amplitude of swing for nonparetic and hemiparetic legs following clinical knee jerks in childhood hemiplegia. Tendon taps were applied to patella tendons of children seated with legs dangling freely at rest. All 12 cases were tested. Traces with evidence of restlessness before and after taps were discarded. A paired comparison of 7/12 cases showed no statistical difference in duration of damped oscillation or amplitude of swing between nonparetic and hemiparetic legs.



249434/437/484

Congenital Right Hemiplegia in 10 year old girl.

Comparison of Right and Left knee jerks and "free fall."

A. Knee jerks. B Legs released from full extension. ↓

Damped Oscillation and amplitude of left leg is approximately twice that of the right.

Q: quadriceps, H: Hamstrings, Gon: goniometer. Paper speed 1.5 cm/sec

Figure 5.7.2 Wartenberg's pendulum test following the knee jerk and "leg drop."

5.8 Discussion of findings.

5.8.1 Methodological pitfalls

These studies demonstrate little excess reflex excitability of the thigh muscles in hemiplegic cerebral palsy using either sudden ramp or continuous, rhythmic, sinusoidal methods of stretch. The hamstrings show greater velocity sensitivity than the quadriceps when subjected to stretch at different initial muscle lengths. However, with the exception of the hemiparetic hamstrings close to full knee extension, hemiplegic muscles are not more excitable than the nonparetic muscles when compared length for length. An assessment of traditional knee jerks using the Wartenberg's pendulum test failed to demonstrate differences in excitability to a strongly phasic stimulus between sides. This implies a similar processing by the spinal cord of afferent input from the muscle spindles: for instance, a similar degree of presynaptic inhibition (see 5.8.3, below and section 2.3.5.).

There may be several explanations for these results each of which is examined in turn.

1. Hemiparetic muscles are more excitable but the method of reflex EMG detection was too insensitive. Since the skin and underlying tissues act as low frequency (high pass) filters for the EMG signal (Basmajian and de Luca 1985), some of the high frequency EMG activity may have been "missed" using surface electrodes and the SLE 800 recorder. There is a long list of disadvantages regarding use of needle or wire electrodes compared with surface EMG, including pain and discomfort, noisy signals during imposed movements and the fact that only muscles within 100μm distance of the electrode are recorded are the principal drawbacks. The advantage of wire recordings is that they minimise the effects of cross-talk between agonist-antagonist muscle groups. The sensitivity of 10 μV/mm appears reasonable for detecting low amplitude signals. Very small signals recorded during a study of Wartenberg's pendulum test in the same children were sufficient to alter the profile of damped oscillations at the knee (Figure 5.7.1).

2. The method of manually stretching the muscles may have failed to elicit a reflex response but both ramp stretching and sinusoidal oscillation are extensions of the bedside clinical method for assessing tone. In this study, the position of the knee and the electrical changes of the muscle were recorded without discomfort to the child and the approach to stretching was standardised. It is inevitable that small variations occurred from muscle to

muscle or subject to subject which might have been eliminated by mechanical stretching techniques using a torque motor, but the advantages of a clinical method capable of being used in any setting would be lost.

3. It could be claimed that the principle of studying muscles from rest is unphysiological whereas assessing partially active muscles might have shown more reflex excitability. Indeed, much of the selective bias in this study involved excluding recordings thought to be 'contaminated' by background EMG of voluntary and involuntary origin. Many studies, relying on minimally contracting muscles (usually 10-20% of maximum voluntary contraction) have demonstrated how differently the motor system can behave, depending on the extent of muscle pre-activation. Cutaneomuscular reflexes (Evans *et al* ,1989;1990 and 1991), long latency stretch reflexes (Berardelli, 1982 and 1983) and magnetic brain stimulation (O'Sullivan *et al* ,1991) all require active muscles before they can be demonstrated and cannot be elicited at rest. However, one of the difficulties with such approaches lies in standardising the level of baseline muscle activity. In calculating 10-20% of maximum voluntary contraction, there is an assumption relating to maximal effort, which is liable to vary considerably between subjects. Such techniques also require a method of signal averaging over many cycles so that the random background activity is averaged out, while the recurring reflex event is summated .

At the outset of this study it was anticipated that the resistance to slow stretch would be a combination of reflex contraction and biomechanical resistance. Provided the children were completely relaxed, there was little reflex EMG whatever the method of muscle stretch. It was easy to disregard sequences in which continuous EMG activity preceded stretch. Voluntary or involuntary activity contaminating an EMG sequence could usually be distinguished from true reflex activity if during a sinusoidal stretch sequence the EMG was originally silent and became briefly active (fig 5.3.5.2A). Actively contracting muscles from either the nonparetic or hemiparetic limbs all showed an augmented reflex response to stretch (fig. 5.3.5.2A) with a corresponding unloading response producing a muscle silent period (Angel 1973; Struppler *et al*, 1973) which became undetectable when muscle pre-activation ceased.

4. Burke *et al* (1971) and Rack*et al* (1984) reported fatigue in their studies of sinusoidal stretch in the thigh and calf respectively. From these records, there were no obvious patterns

consistent with fatigue: on the contrary, the subjects were usually extremely relaxed!

5. It has been demonstrated that the stretch-evoked cortical potential diminishes with an increasing frequency of presentation of the stretch stimulus of wrist extensors (Rothwell *et al*, 1987). This results in a long latency EMG response of smaller amplitude as the stimulus frequency increases from 0.1 Hz to 1Hz. By definition, monosynaptic, short latency responses are unaffected by this long-latency habituation.

6. There is always the possibility of a larger study producing statistically significant results that are clinically irrelevant. This study involved repeated measurements on four muscles in each subject at three different muscle lengths using two distinct methods of stretch in 13 subjects.

All the possible methodological pitfalls mentioned above would still need to explain why the hemiparetic and nonparetic reflex EMG responses to stretch were essentially similar .

5.8.2 Biomechanical versus reflex resistance.

This relatively electrically silent resistance to stretch at rest suggests that the resistance felt must be biomechanical in nature, even when the knee joint is close to full extension and the quadriceps and hamstrings are at their most excitable. This implies that the increased resistance or "stiffness" may be due to peripheral muscular transformation of either the contractile elements of muscle or adjacent non-contractile elements (see sections 2 and 4 for further discussion). Evidence for a lack of reflex activity in resting "spastic" muscles was confirmed by Foley in 1961. He demonstrated that the velocity-dependent hypertonus in resting normal and spastic adults and children with spastic diplegia was independent of reflex EMG activity. He showed very clearly the phenomenon of "stress relaxation" which is the gradual reduction in torque required to maintain a given muscle length over time. He also showed that physical therapy involving limb and trunk stretching could significantly reduce muscle tone whereas complete peripheral nerve blocks had no effect.

5.8.3 Why should children be different from adults?

There are several possible reasons why the effects of central motor injury in adults differ from those which arise from congenital injuries or those sustained in early infancy. Herman (1970) showed differences in the reflex and the viscoelastic properties of muscle in relation to duration and severity of the stroke symptoms. Likewise, Thilmann *et al* (1991) have demonstrated that maximum EMG activity occurs between the first and second month

following a stroke in the biceps muscles of 19 hemiparetic adults, declining thereafter in the first year. This evidence suggests that reflex excitability diminishes with time or "burns out" and this may equally apply to children though in most cases the clinical pattern of reflexes were never particularly brisk. Evidence is emerging that the growing and developing brain responds to damage in some cases, by the fast -conducting cortical motor units of one hemisphere supplying bilateral lower motor neurone pools (Carr *et al*, 1991; 1993) and producing mirror movements in some cases. In an adult who had undergone a left hemispherectomy for intractable seizures secondary to a congenital porencephalic cyst, Cohen *et al*, 1991 have demonstrated activation of the biceps and deltoid of the hemiparetic arm when the surviving cortex was magnetically stimulated 2cm anterolateral to the point of activation of the nonparetic side. Voluntary movements of the affected limb produced an increase in cerebral blood flow in an area 1.4 cm anterior to that produced by movement of the nonparetic arm and also produced a bereitschaftspotential (pre-movement potential) in the surviving ipsilateral hemisphere. This cerebral plasticity appears to be confined to motor reorganisation since no sensory evoked potentials to median nerve stimulation could be elicited from the hemiparetic limb. This motor reorganisation may prevent the loss of presynaptic inhibition which results from damage to supraspinal structures in adults. Evidence supporting plastic reorganisation of the brain comes from the observation that clinical signs, function and disability appear to bear little relation to the size and distribution of the lesion on CT imaging (Wiklund and Uvebrant, 1991) in hemiplegic cerebral palsy. Another possible palliating factor stems from the fact that the principal cause of congenital hemiplegia appear to be antenatal and the intrauterine environment may provide support in maintaining homeostasis even in the presence of a foetal stroke, offsetting secondary hypoxic-ischaemic injuries.

Of the 18 cases studied by Burke *et al*, 1971, only one case had spasticity of cerebral origin attributed to a parasagital meningioma and in 5/18 of the cases it was not possible to elicit a sinusoidal reflex threshold. These adult cases appeared to have much lower reflex frequency thresholds of the order of 0.5 Hz compared with the children in our study in whom it was not possible to elicit a reflex at frequencies of less than 1 Hz. A partial explanation of this is that the adults in the Burke study had mainly spinal pathologies, including multiple sclerosis in 6/18 cases. Tatton *et al* (1985) report that only 70% of patients with increased

tone, as a sequel to cortical or internal capsular vascular lesions, show increased short-latency EMG activity. Tatton's findings (1985), and those of Burke and colleagues (1971) support the notion that in at least 30 % of adult strokes, non-electrical hypertonus is present.

5.9 Summary.

The quadriceps and hamstring muscles of thirteen children with hemiplegic cerebral palsy have been studied using ramp and sinusoidal stretches at three different muscle lengths. Overall, the hamstring muscles showed greater velocity sensitivity than the quadriceps with the hamstrings having the most sensitive reflex velocity and frequency thresholds close to maximum knee extension. At this position, the hemiparetic hamstrings alone, showed a weakly significant lower reflex frequency threshold compared with nonparetic muscles. For all other muscle lengths, nonparetic and hemiparetic muscles displayed similar reflex thresholds when subjected to sudden, discontinuous ramp or repetitive, rhythmical, sinusoidal stretches. The number of muscles for which a reflex threshold could be established declined progressively as the angle at the knee joint approached 90° of flexion. Muscles for which no reflex threshold could be demonstrated were subjected to similar velocities and frequencies of stretch as those in which a reflex was obtained.

5.10 Conclusion.

In adult populations, one third of the patients with hemiparesis do not exhibit excessive reflex excitability; such excitability appears to wane following the first year after the injury, cerebral damage produces intrinsically less excitability than spinal injuries. Children may exhibit less reflex excitability because of the age of the injury, biomechanical transformation of muscle and plastic reorganisation of corticospinal pathways.The implications for management are that if the resistance to stretch is not in the main due to a velocity-dependent electrical reflex, drugs and neurosurgical management specifically aimed at reducing spasticity is clearly inappropriate. While the treatment of hemiplegic children tends to be conservative, this cannot be said for the radical treatment options offered to diplegic children (Peacock and Staudt, 1990; Landau and Hunt, 1990). Systematic evaluation of reflex excitability in individual cases ought to help to establish a more realistic perspective of its functional significance not only in the hemiplegias but in other types of cerebral palsy.

6. Distal Lower Limb Reflex Excitability and Function.

6.1. Background.

In section 5, doubt has been raised regarding an absolute increase in reflex excitability of the quadriceps and hamstrings muscles in hemiplegic cerebral palsy. Although both the hemiparetic and nonparetic quadriceps and hamstring muscles obeyed the same principles of maximum excitability (velocity-dependence) close to full knee extension, and reflex excitability was extinguished close to 90° of knee flexion in the majority of muscles, no clear differences in reflex velocity or frequency thresholds could be established between the nonparetic and hemiparetic muscles.

In this section, a method of measuring the stretch reflexes of the gastrocnemius-soleus muscles acting across the ankle joint is described for hemiplegic children as a formal extension of the clinical examination. The aim was to measure the reflex electromyogram (EMG) elicited, not the resistance felt (which would require other techniques). The velocity-dependence of the reflex EMG in plantarflexor muscles is demonstrated. The reflex excitability on the nonparetic and hemiparetic sides is compared, and the potential clinical applications of assaying stretch reflexes are discussed in the wider context of all the cerebral palsies.

6.2 Methods.

6.2.1 Patients

A convenience sample of 14 children with congenital hemiplegia of mean age 9.3 years (SD 2.2, 6.2-12.2) were examined under controlled conditions. The clinical background of each child is summarised in table 6.21 which includes a brief description of the intervening therapy to which the child had been exposed. All the children had received "eclectic" physiotherapy (PT) in the form of slow, passive stretching administered by the parents and under 6-8 weekly supervision of a professional physiotherapist from the time of diagnosis of the hemiparesis. A further 9/14 had been using an ankle-foot-orthosis (AFO) for between 2 and 8 years depending on age and additionally, 4 of these also had recourse to a night splint, for between 2-5 years prior to enrolment. One child had only had a night splint with PT. Five out of 14 children proceeded to serial ('inhibitory') casting either as one-off procedures over 4-6 weeks in 3/5 cases, twice in 1/5 cases or yearly in 1/5 cases. One child subsequently underwent heelcord tenotomy 3 years prior to the present assessment. In each of these cases, table 6.2.1 gives the time interval between the onset of a given therapy and the time of reflex assessment.

Table 6.2.1 Details of hemiparetic children.
from Lin, Brown and Brotherstone, 1992, by permission.

Details of hemiplegic children by case number, age, sex, side of hemiplegia, congenital-perinatal or acquired aetiology, associated childhood seizures, special educational needs, gait grade, muscle tone and treatment. n = 14.

Case	Age (yrs)[1]	Sex	Aetiology[2]		Side of hemiplegia	Seizures	Special education	Gait grade[3]	Tone				Treatment[4]				
			Congenital	Perinatal					Phasic spasticity	Tonic spasticity	Hemi-dystonia	Normal tone	Physio-therapy	AFO	Night splint	Teno-tomy	Inhib. cast.
2	11·0	M	IUGR		R	+	+	4	+	+	–	–	10	6		3	6,4*
4	11·7	M	IUGR		R			4	–	+	–	–	7	2	3		Yearly
6	9·3	F	Silent		R			1	+	+	–	–	8				3
8	10·2	F		Asphyxia	R	+	+	2	–	–	–	+	9	8			
10	6·4	M	Silent		R			1	–	+	–	–	5				
11	6·7	M	Silent		R	+	+	4	–	+	–	–	8	1	4		
12	11·3	F	Preterm		R			2	+	+	–	–	10				
14	6·7	M	Silent		R			3	–	+	–	–	6	4			
15	11·1	M	Silent		L			2	–	+	–	–	2	2			
16	8·2	M	Preterm		R			4	+	+	–	–	6	2			
17	6·2	M	Silent		R			4	+	+	–	–	5	4·5			4
18	12·0	M	IUGR		L			3	+	+	–	–	11		2		
22	7·6	M		Asphyxia	L			3	+	+	–	–	7	5	5		1
23	12·2	F	Silent		L			1	–	–	+	–	11				

[1]Mean (SD) = 9·3 (2·2).
[2]Silent = presumed silent infarct; IUGR = intra-uterine growth retardation; there was none with acquired hemiplegia.
[3]Gait score: 1 = heel strike, 2 = plantar strike, 3 = toe-heel strike, 4 = toe strike.
[4]Years intervening between treatment and assessment; AFO = ankle-foot orthosis; inhib. cast. = inhibitory casting.
*Six and four years before assessment.

6.2.2 Goniometry.

The ankle hind-foot displacement was measured in degrees with a Penny and Giles® flexible goniometer as shown in figure 6.2.2a-d. Displacement over time was monitored on an eight-channel SLE 800 EEG polygraph recorder with a bandwidth of 0.5-150Hz using a paper speed of 1.5cm/s and a display gain of 10μV/mm . With the foot held in slight inversion at the ankle, the examiner applied a displacement of 20°-40° amplitude from maximum plantarflexion (6.2.2a), through the position of resting plantarflexion (RPF, fig 6.2.2b) and the right angle position (neutral, fig 6.2.2c) of the ankle joint, into dorsiflexion beyond neutral (fig.6.2.2d). With the knee fully extended, these manoeuvres produced stretches of the soleus and gastrocnemius muscles acting through the Achilles tendon. The same manoeuvre in reverse stretched the Tibialis anterior muscle acting across the ankle joint (figure 6.2.2d-a).

6.2.3 Manual torque stretches.

The children lay comfortably in the prone position with the feet over the end of a couch. Stretches were applied by the examiner, without recourse to a torque motor. This has the advantage that the stretches merely represented an extension of normal clinical practice with records of joint position and muscle activity.

The stretch reflex can be specified if the position and speed of stretch of the muscle are known. How the muscle is stretched may also affect reflexes. The Tibialis Anterior (TA) and Gastrocnemius-Soleus (GS) muscle stretch reflexes were studied using two types of stretches.

1. Sudden ramp or discontinuous stretches at increasing angular stretch velocities were used to test the muscles under conditions which mimic sudden displacements such as landing on one's feet from a height.

2. Rhythmic, continuous, sinusoidal stretches at increasing stretch frequencies were used because they represent the cyclical motions of limbs about joints most commonly encountered in physiological movements and also because they reduce the effects of acceleration and deceleration (abrupt changes in direction) occurring between dorsiflexion and plantarflexion.

These two types of stretch broadly represent the two main types of perturbation experienced at the ankle in every-day life.

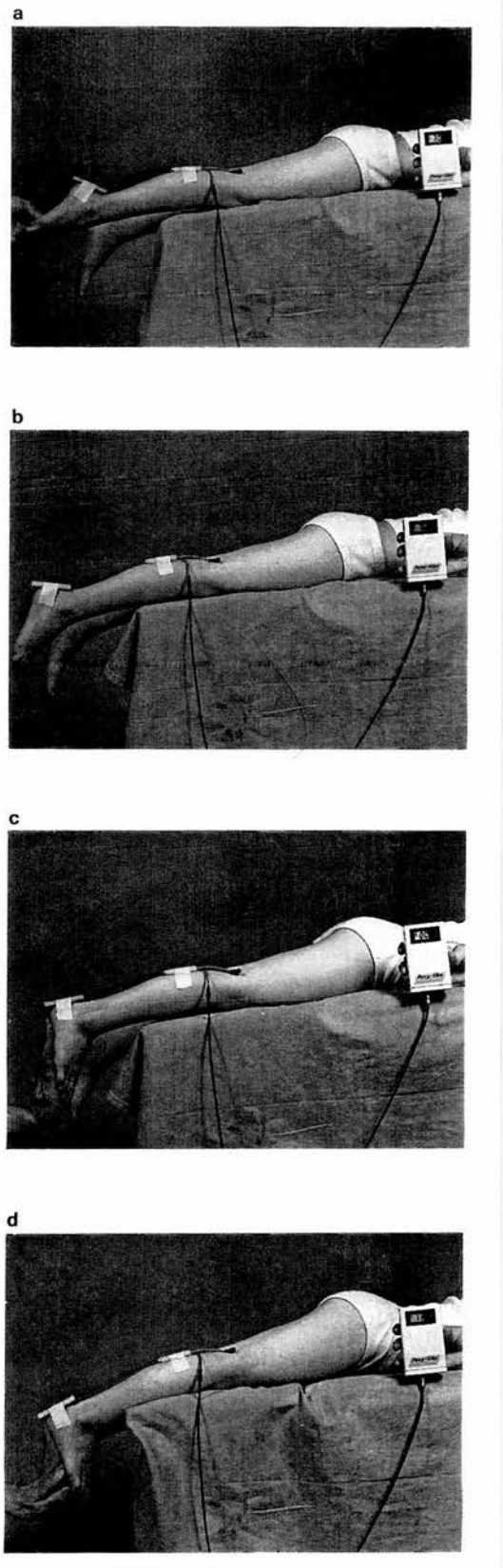


Figure 6.2.2a-d
Ankle goniometry.
A flexible Penny and Giles twin-axis goniometer is attached to the back of the leg in the mid-line with the distal end taped to the heel.
a. max. passive plantarflexion,
b. resting plantarflexion,
c 'neutral angle (90°)and
d. max. passive dorsiflexion.
Passive joint range=d. minus a.
Effective joint range=d. minus b.
Lin, Brown and Brotherstone, 1994b. By Permission

In order to stretch a muscle over a given range at a variety of speeds, the examiner varied the applied torque. Absolute limitations to speed of stretch being a combination of the inertial characteristics of the child's foot and the examiner's hand and arm (the force of stretch increasing with the square of the frequency of stretch, see Section 9 and Walsh, 1992, p37) and the combined mechanisms producing stiffness of the muscles resisting stretch (see section 2: The physiology of muscle tone). The torques applied will in many cases have exceeded several Nm.

6.2.4 Voluntary, involuntary and reflex electromyography (EMG).

Bipolar surface electrodes (Medicotest "Blue Sensor") were placed parallel to the shaft of the leg in the midline, with an inter-electrode distance of 6-10 cm over the TA and GS muscles, depending on the size of the child. The EMG was recorded using an eight-channel SLE 800 EEG polygraph recorder with a bandwidth of 0.5-150Hz using a paper speed of 1.5cm/s and a display gain of 10μV/mm.

Stretches were imposed on initially resting muscles. All polygraphic recordings show a time signal, dorsiflexor (TA) EMG in micro Volts (μV), ankle displacement (GON) in degrees (°) and the plantarflexor (G-S) EMG in micro Volts (μV). By convention dorsiflexion (DF) is upwards, indicated by an arrow, and plantarflexion (PF) downwards for sinusoidal and ramp stretches

Measurements of resting plantarflexion (RPF) and the passive joint ranges were obtained together with the maximum voluntary isometric EMG of the plantarflexor and dorsiflexor muscle groups. The maximum speed of voluntary alternating plantarflexion - dorsiflexion (PF/DF) at the ankle was recorded in Herz (Hz) as a measure of selective motor control or 'ankle dexterity' (AD) together with the amplitude of voluntary movement in degrees.

Ankle jerks were also recorded along with the number of beats of ankle clonus elicited.

6.2.5 Analyses.

Traces were analysed qualitatively according to the type of test with regard to recognisable EMG patterns and the presence or absence of background activity. The results for hemiparetic and nonparetic limbs are plotted against either the velocity of ramp stretches, in degrees/second (°/s), or the frequency of sinusoidal stretches in Herz (Hz).

In most cases, the reflex frequency and reflex velocity thresholds and gains, respectively, could be calculated from the regression equation of the reflex EMG-velocity or reflex EMG-frequency plots. Reflex EMG thresholds were derived from the intercept of a linear regression equation of the EMG amplitude plotted against either frequency or velocity of stretch. The slope of the regression equations defined the reflex EMG gain. Since the data for each limb was gathered over time, variations in mental state due to the influence of non-specific stimuli were averaged over the same period and the regression equations for each limb and each mode of stretch were thus specified in terms of reflex thresholds and gains which represent the "reflex excitability" of the limbs at that time.

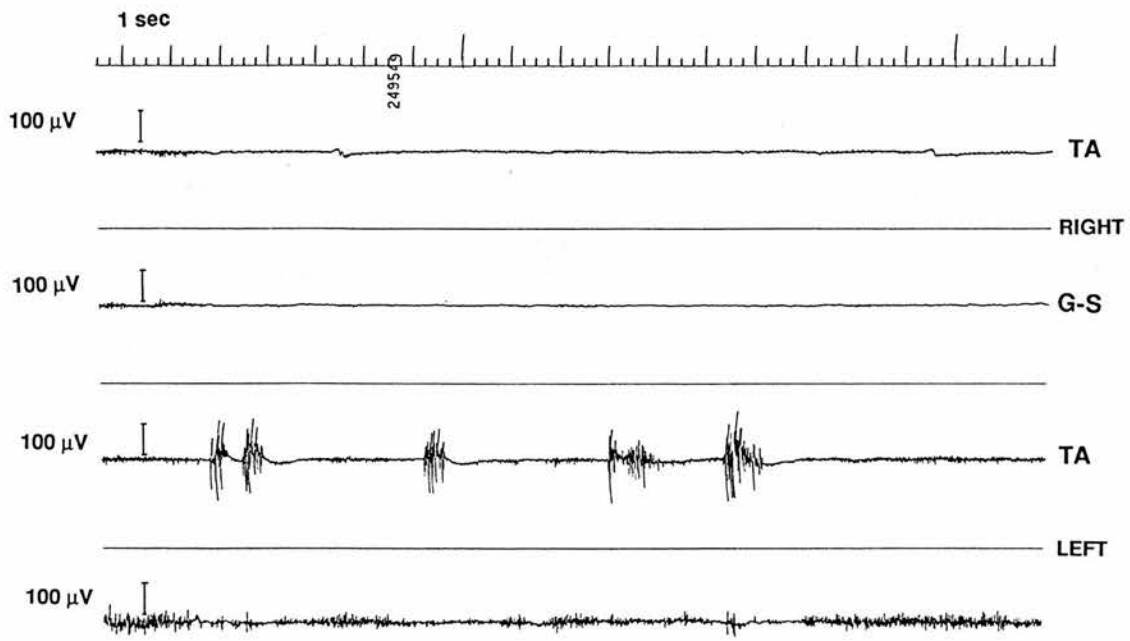
Data is tabulated in the form of 99% confidence intervals. Hemiparetic and nonparetic reflex thresholds, reflex gains and ankle dexterity measures were compared for each case using paired "t" tests as were the passive ankle joint ranges obtained.

6.3 Results.

6.3.1 Qualitative joint angle and EMG analysis in standing.

Figure 6.3.1(i) indicates the tibialis anterior (TA) and gastrocnemius-soleus (GS) EMG recordings during double-limb standing for a 10 year old with a congenital right hemiparesis. Recordings from the hemiparetic right leg show little or no EMG activity, whereas the left leg shows bursts of discharges corresponding to a gentle swaying motion. The TA muscles being active when the body is swaying backwards, the G-S muscles are active during forward sway. There is no perceptible joint angle change. Figure 6.3.1 (ii) compares similar standing EMG patterns from the TA and G-S muscles of a further two children: a 12 year old boy with a congenital left hemiparesis and a 6.5 year old boy with a right hemiparesis. In both cases the hemiparetic limb shows only low-grade activity in standing compared with the nonparetic side in which the TA muscles are particularly active.

The standing EMG of a 12 year-old girl with a congenital left hemidystonia (figure 6.3.1 (iii)) shows that the non-dystonic limb is the most active, particularly the TA muscles, with the dystonic side exhibiting only low-grade EMG activity. When the same subject begins to walk, figure 6.3.1 (iv), the hemidystonic left leg shows evidence of greater co-contracton than the non-dystonic side, but again the TA muscles appear most active. In standing, the dystonic and non-dystonic hemiparetic limbs behave similarly with an overall reduced motor output for muscles acting across the ankle joint. Nonparetic TA muscles side are most active.



249549

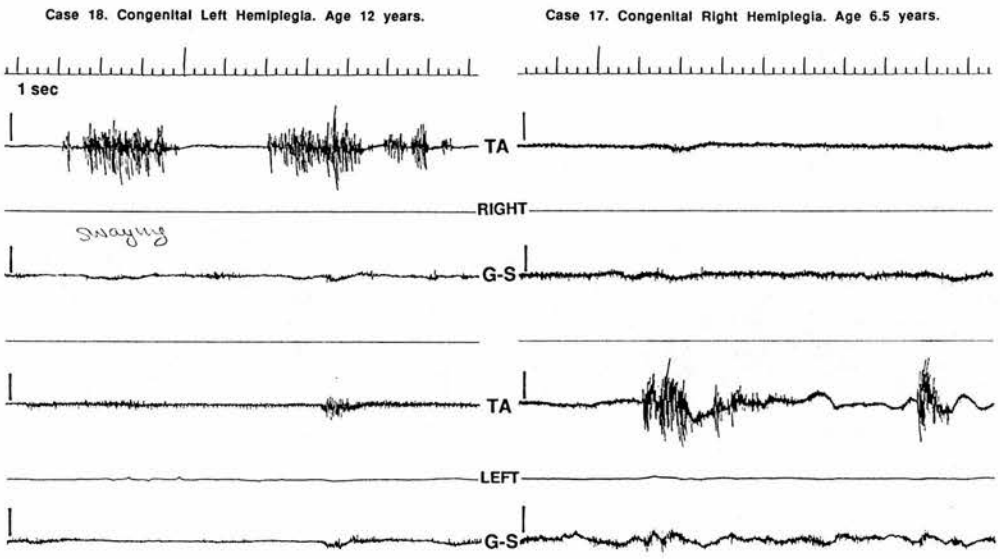
Congenital Right Hemiplegia in 10 year old girl.

Standing almost at rest. Bursts of Left TA activity correspond to swaying.

By comparison the abnormal Right sideis virtually silent.

TA: Tibialis anterior. G-S: Gastrocnemius-Soleus. GON: goniometer

Figure 6.3.1.(i) Standing leg muscle EMG in a case of right hemiparesis.

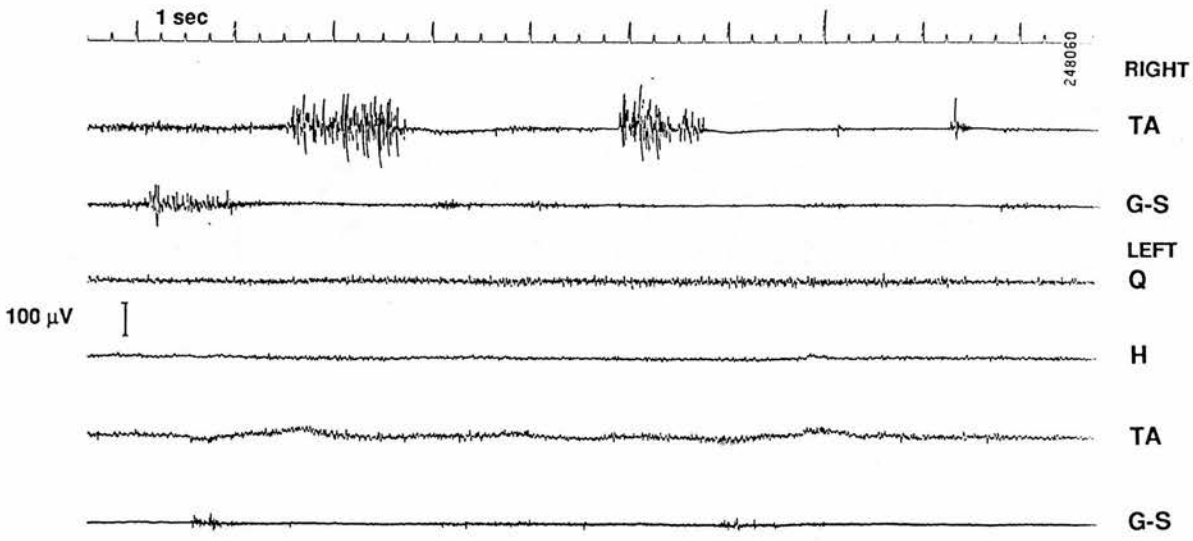


Comparison of EMG during standing in normal and affected distal limbs in hemiplegia.

Note activity in TA of "normal side" during sway .

TA: Tibialis Anterior EMG, G-S: Gastrocnemius-Soleus EMG, GON: Goniometer.
Calibration bars: EMG 100μV, goniometer 20°, time 1 sec.

Figure 6.3.1 (ii). Standing leg muscle EMG in a case of left and right hemiparesis.



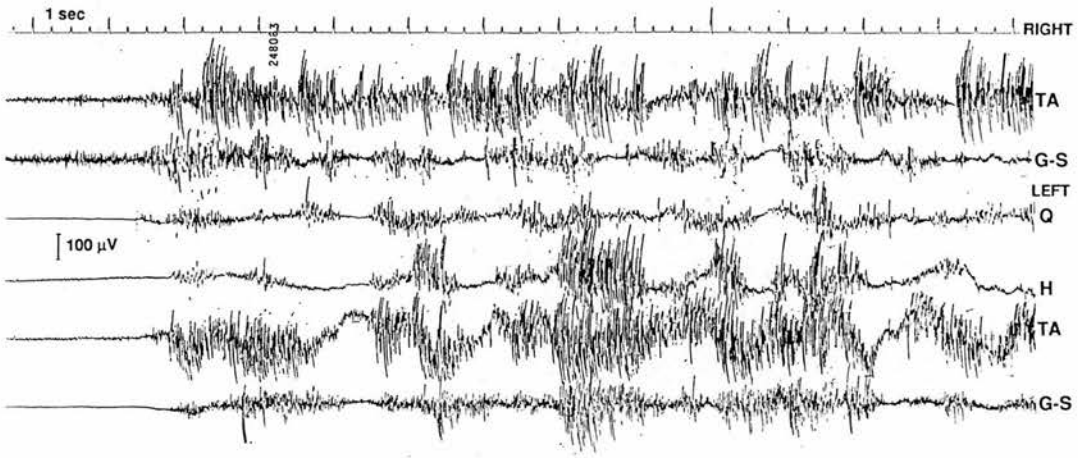
248060

Congenital left hemidystonia in 12 year old girl.

Standing: minimal EMG activity results from sway and fidgeting in right and left legs.

TA: Tibialis anterior, G - S Gastrocnemius-Soleus, Q: quadriceps, H: Hamstrings, GON: goniometer. Paper speed 1.5 cm/sec.

Figure 6.3.1 (iii) Standing leg muscle EMG in left hemidystonia



248063

Congenital left hemidystonia in 12 year old girl.

Standing followed by walking. Note virtually silent EMG in dystonic Left leg prior to walking.

TA: Tibialis anterior, G - S Gastrocnemius-Soleus, Q: quadriceps, H: Hamstrings, GON: goniometer. Paper speed 1.5 cm/sec.

Figure 6.3.1 (iv) Standing followed by walking EMG in a case of left hemidystonia.

6.3.2 Isometric contractions at the ankle.

The EMG and goniometer output during isometric plantarflexion and dorsiflexion are shown in figure 6.3.2 (i)A for the hemiparetic and B the nonparetic limbs of a 10 year-old girl with a congenital right hemiparesis. The output is largest for TA and G-S of the non-paretic limb. Similar findings are evident in a 6.5 year-old boy with a right hemiparesis, figure 6.3.2 (ii), though there is greater evidence of co-contraction than in figure 6.3.2 (i).

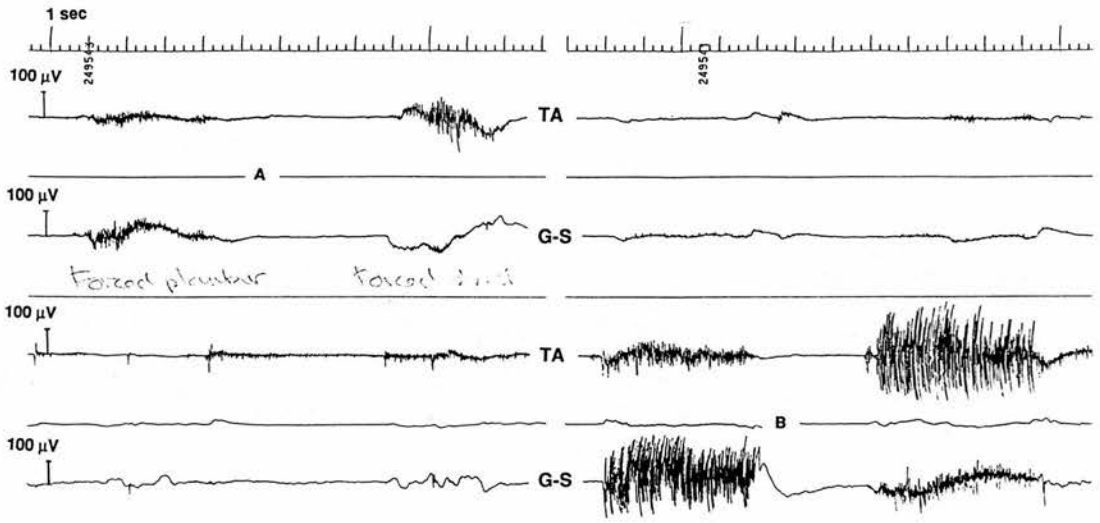
6.3.3 Voluntary alternating movements at the ankle.

The children were asked to waggle their feet up and down as fast as possible. Figure 6.3.3 (i) illustrates this for the nonparetic limb of a 10 year-old girl with a congenital right hemiparesis which can perform high frequency, high amplitude motions, figure 6.3.3.(i)A which displays high amplitude TA and G-S activity. Figure 6.3.3.(i)B. shows high frequency, low amplitude motions in the same nonparetic limb, but now the TA muscle EMG discharge amplitude is much reduced indicating the reduced effort of the lower amplitude motions. The phasic TA EMG discharges in figure 6.3.3(i)B also demonstrate EMG silence at peak dorsiflexion, the peak TA EMG discharge occurring near to maximum plantarflexion, ie at 180° in phase advance of peak dorsiflexion (see section 5.3.5 and figure 5.3.5.1a for a full discussion of the phase relations between EMG and joint angular displacement).

Figure 6.3.3 (ii) shows attempted alternating movements at the ankles (A) and toes (T) of the the hemiparetic (top traces) and nonparetic (bottom traces) limbs of the same subject as for figure 6.3.3 (i). The hemiparetic alternating movements are of low amplitude and low frequency which tallies with a low amplitude tonic EMG output and evidence of co-contraction between TA and G-S muscles.

The EMG and goniometer output during alternating movements at the ankle for a 12 year-old boy with a left hemiparesis (case 18) and a 6.5 year-old boy with a right hemiparesis are shown in figure 6.3.3 (iii) with similar results to the findings in figure 6.3.3 (ii). The reduced, poorly differentiated output from the muscles of the affected limb, particularly the TA muscles, appears universal. For both hemiparetic limbs there is a constant, low grade background EMG discharge throughout the period of attempted motion.

Figure 6.3.3 (iv) depicts alternating movements in a 12 year-old girl with a left hemidystonia: the affected limb movements are coarse and poorly modulated.



249543/541

Congenital Right Hemiplegia in 10 year old girl.

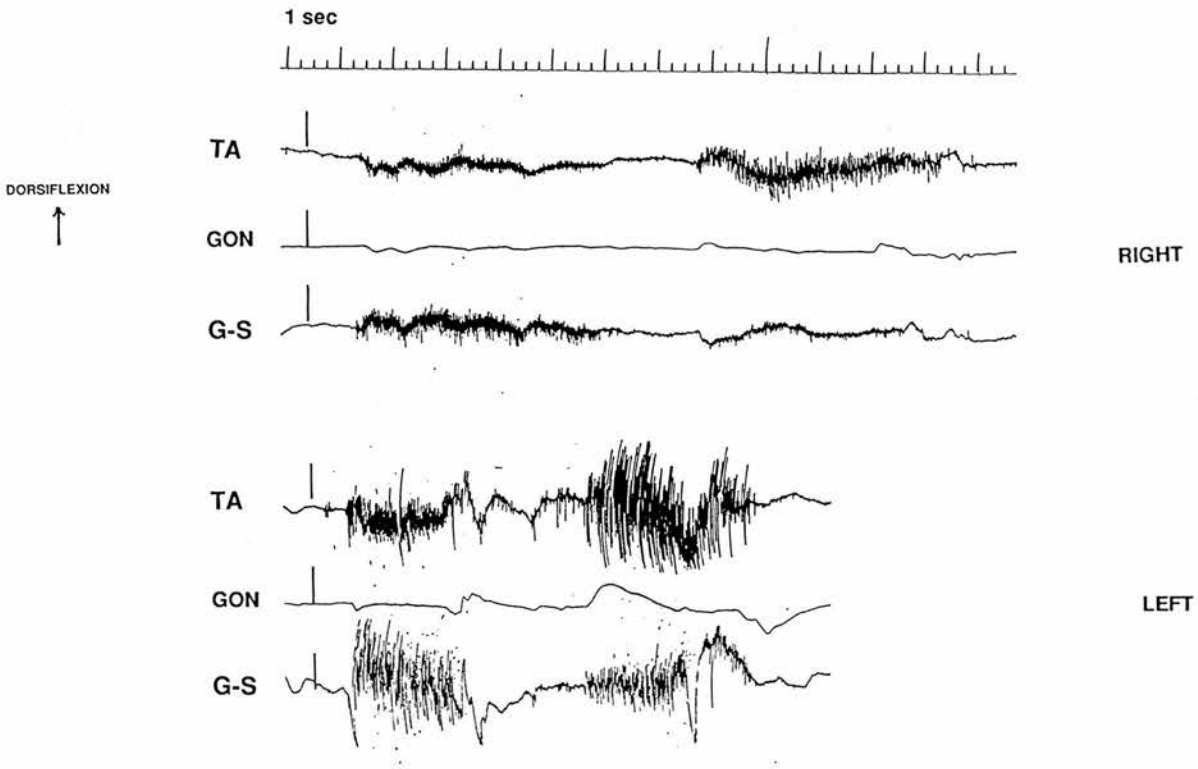
Isometric Right and Left plantarflexion and dorsiflexion.

A. Right side: G-S trace 80 µV, TA trace 140 µV.

B. Left side : G-S trace 300 µV, TA trace 400 µV.

TA: Tibialis anterior. G-S: Gastrocnemius-Soleus. GON: goniometer

Figure 6.3.2 (i) Isometric contractions of TA and G-S muscles.

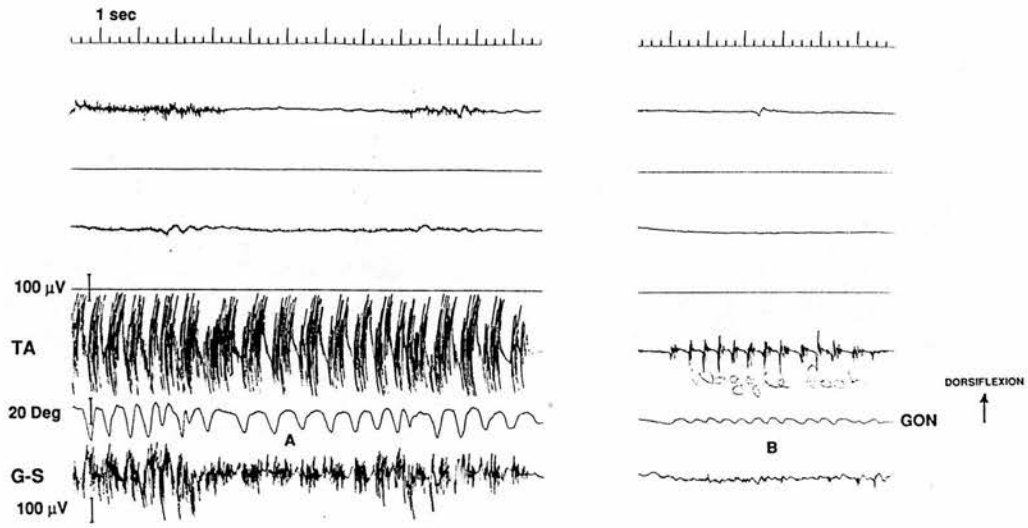


Comparison of isometric plantar and dorsiflexion on "normal" and affected sides.

Case 17. Congenital Right Hemiplegia. Age 6.5.
EMG pattern clearly reduced on paretic side.

TA: Tibialis Anterior EMG, G-S: Gastrocnemius-sSoleus EMG, GON: Goniometer.
Calibration bars: EMG 100μV, goniometer 20°, time 1 sec.

Figure 6.3.2 (ii) Isometric contractions of TA and G-S muscles.

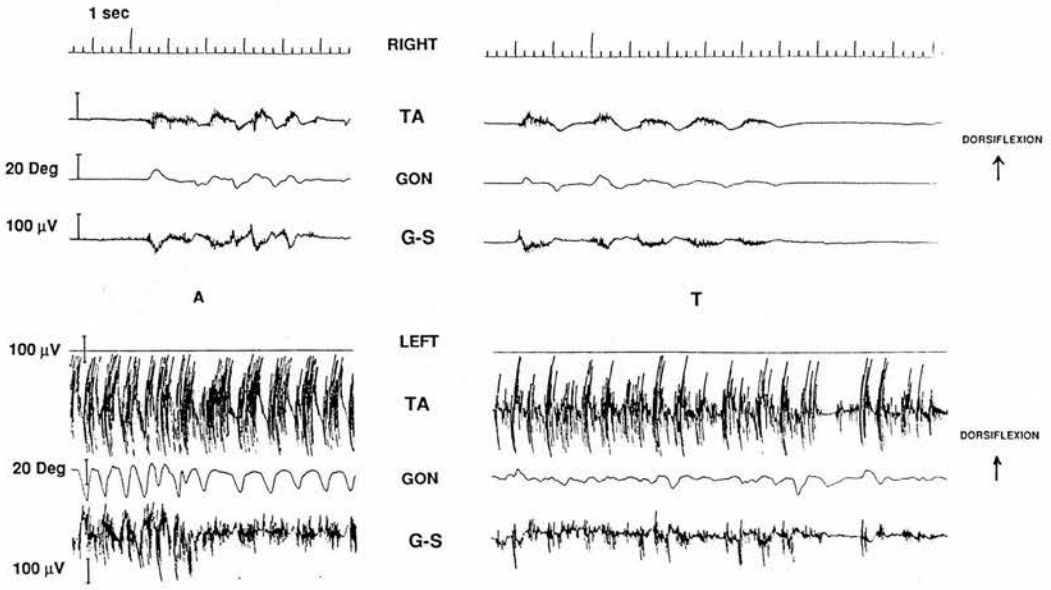


249533/543

Congenital Right Hemiplegia in 10 year old girl,

voluntary Left alternating plantar/dorsiflexion. A.1-2 Hz. for 18-26 deg displacement. B. 1.8 Hz for 4 deg. with mostly active TA. TA: Tibialis anterior. G-S: Gastrocnemius-Soleus. GON: goniometer

Figure 6.3.3 (i) Modulated alternating movements at the ankle of the nonparetic limb.



249513/515/533/534

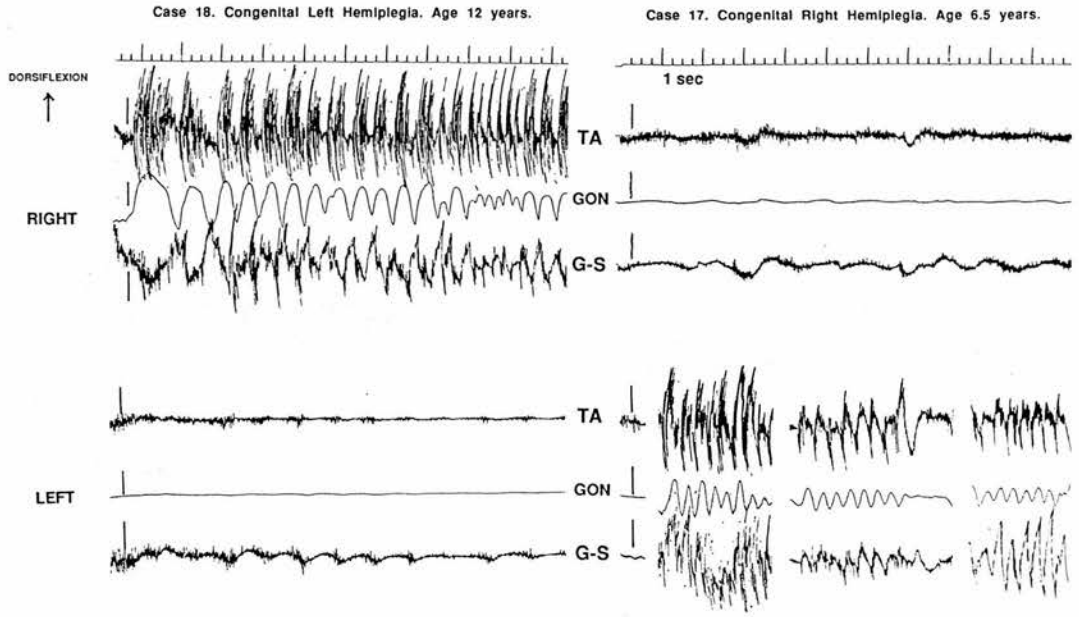
Congenital Right Hemiplegia in 10 year old girl.

Voluntary movement at the ankle and toes of Rght and Left feet.

		Frequency Hz	
		R	L
A	Ankle	1	2
T	Toes	0.5	1

TA: Tibialis anterior. G-S: Gastrocnemius-Soleus. GON: goniometer

Figure 6.3.3.(ii) Comparison of alternating movements at the ankle and toes in hemiplegia.



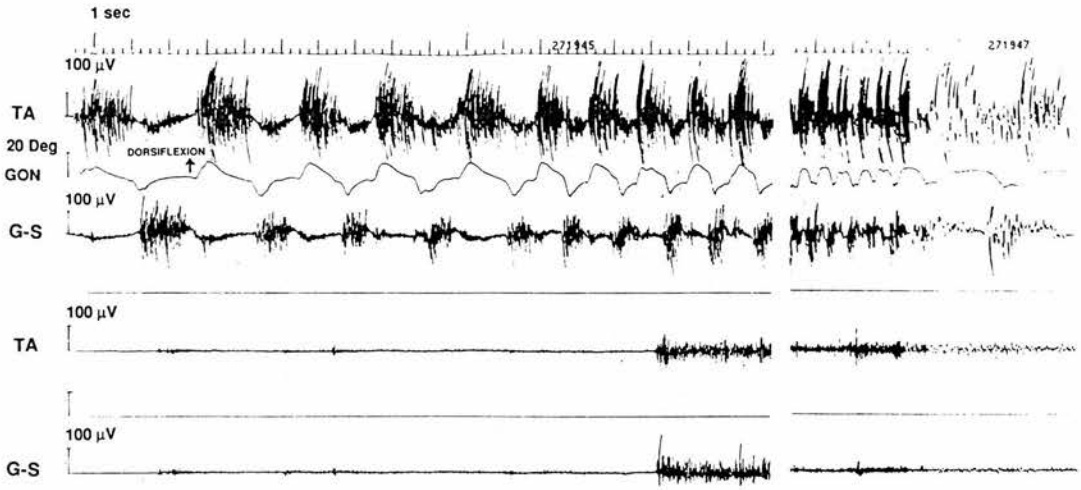
Comparison of motor dexterity at the ankle of normal and affected limbs in hemiplegia.

Patients supine. Isotonic alternating dorsiflexion/plantarflexion at self selected speeds.

Range, speed and smooth control of movement preserved on "normal side".
Motionless affected side. Note low grade co-contraction of affected TA and G-S muscles.

TA: Tibialis Anterior EMG, G-S: Gastrocnemius-Soleus EMG, GON: Goniometer.
Calibration bars: EMG 100μV, goniometer 20°, time 1 sec.

Figure 6.3.3 (iii) Two cases of hemiparetic and nonparetic alternating movements



271945

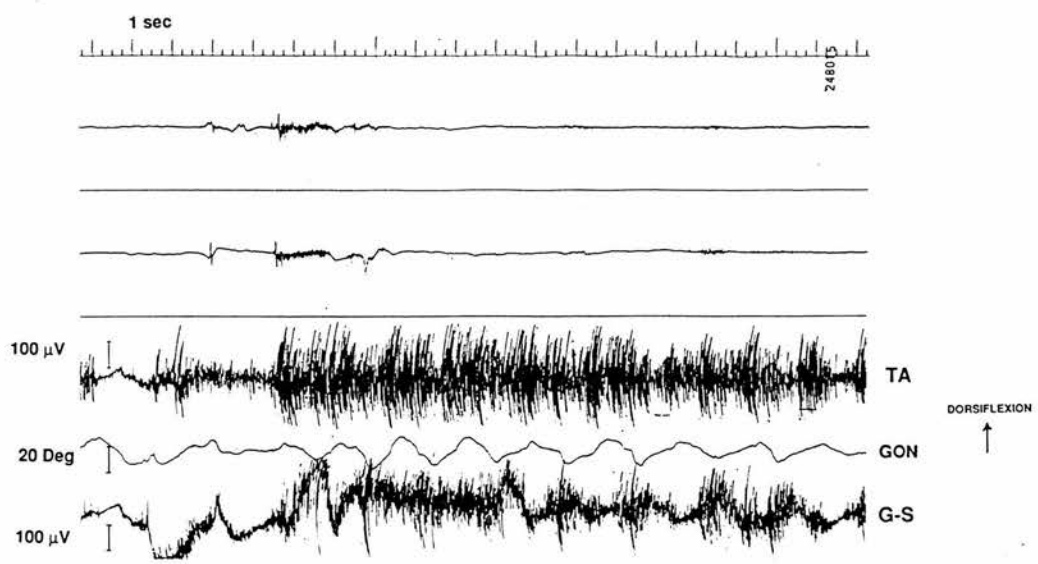
Congenital left hemidystonia in 12 year old girl.

Voluntary alternating plantarflexion / dorsiflexion at right ankle from 1.3 - 6 Hz showing normal triphasic response.

As frequency increases, amplitude diminishes from 26 to 10 degrees peak - peak.

Note dystonic response in Left TA and G-S muscles as patient increases speed of movement of normal limb.

TA: tibialis anterior, G-S: gastrocnemius-soleus, GON: goniometer. Paper speed 1.5 - 6 cm/sec.



248019

Congenital left hemidystonia in 12 year old girl.

Attempted Left ankle voluntary dorsiflexion / plantarflexion.

There is continuous EMG activity with loss of a clear triphasic response.

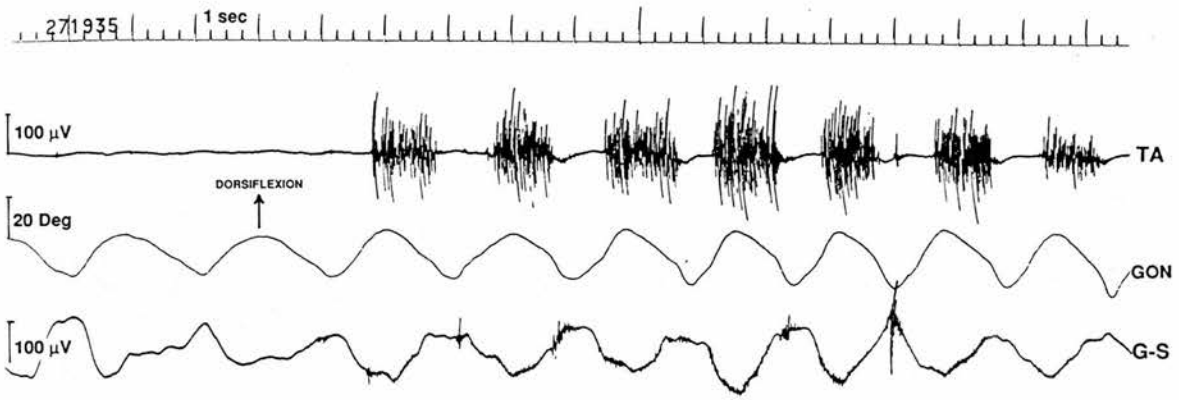
TA: Tibialis anterior, G - S Gastrocnemius-Soleus, GON: goniometer. Paper speed 1.5 cm/sec.

Figure 6.3.3 (iv) Alternating movements at the ankle in left hemidystonia

6.3.4 Passive movements at the ankle joint.

As discussed in section 5.3.3, it is important to interpret the phase relations between EMG activity and angular displacement. In figure 6.3.4(i), passive alternating movements at the ankle (top traces) of the non-dystonic limb are not accompanied by EMG activity for the first three dorsiflexions, then there are 7 bursts of TA muscle activity which delay and distort the plantarflexor phase of the goniometer output. This represents voluntary tonic eccentric TA EMG activity ie the muscle actively resists passive lengthening. As the frequency of the passive motion is increased to 0.8Hz, the bursts of eccentric TA EMG activity increase in amplitude (bottom traces) and are of a shorter duration to match the plantarflexor phase of the passive motion. In both sets of traces, the TA EMG activity is brought on at will.

Figure 6.3.4 (ii), traces from both limbs of the same child with a left hemidystonia are shown. Passive motion at the ankle proceeds unhindered by any voluntary or involuntary muscular activity, but ends with a burst of co-contraction 3 s after the end of passive motion. However, the 'resting' left dystonic limb becomes active two-thirds of the way into the passive stretches of the non-dystonic limb, indicating restlessness. These sequences illustrate purposeful and non-purposeful EMG activity during passive stretches.

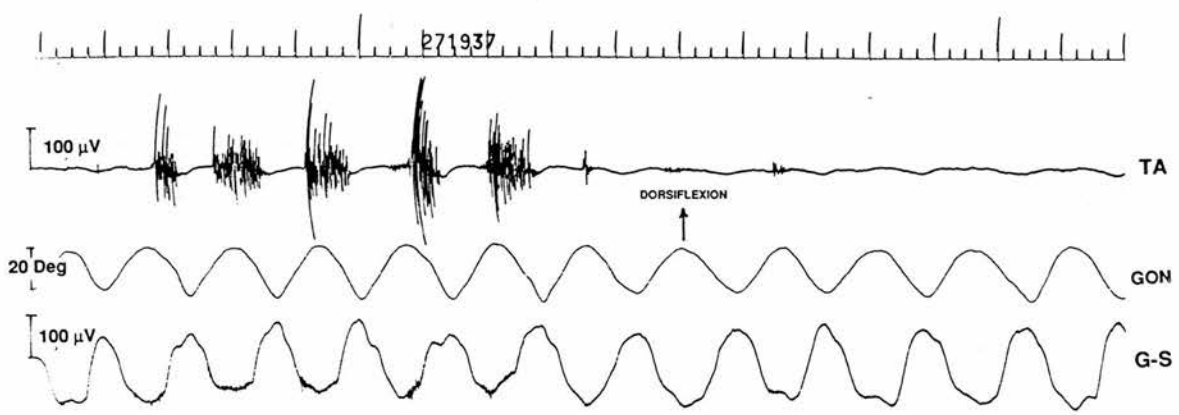


271935

Congenital left hemidystonia in 12 year old girl.

Passive Right ankle dorsiflexion / plantarflexion at 0.45 - 0.6 Hz through a range of 22 degrees.

Tibialis anterior and Gastrocnemius-Soleus initially silent for 2 cycles followed by voluntary dorsiflexion with bursts of tibialis anerior activity. TA: Tibialis anterior, G - S Gastrocnemius-Soleus, GON: goniometer. Paper speed 1.5 cm/sec.



271937

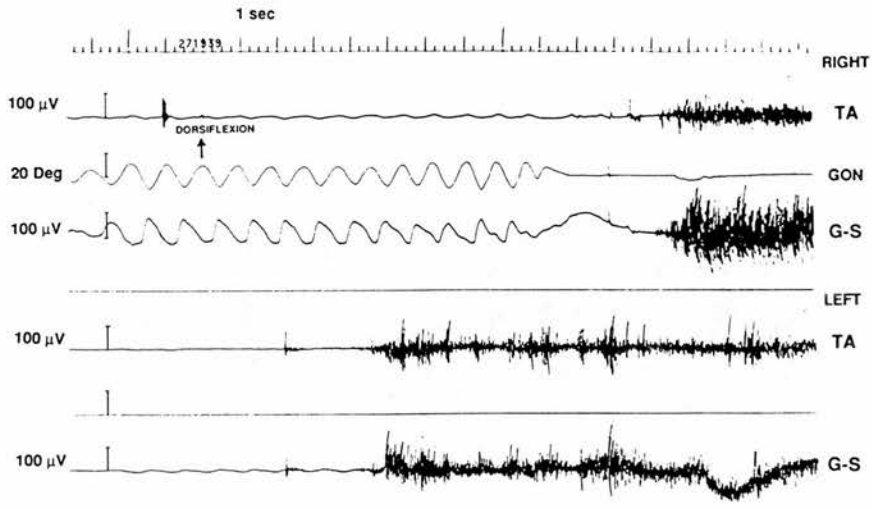
Congenital left hemidystonia in 12 year old girl.

Passive Right ankle dorsiflexion / plantarflexion at 0.8 - 0.6 Hz through a range of 26 degrees.

6 cycles of voluntary dorsiflexion noted followed by muscle silence.

TA: Tibialis anterior, G - S Gastrocnemius-Soleus, GON: goniometer. Paper speed 1.5 cm/sec.

Figure 6.3.4 (i) Eccentric tonic TA EMG activity during passive plantarflexion.(see text)



271939

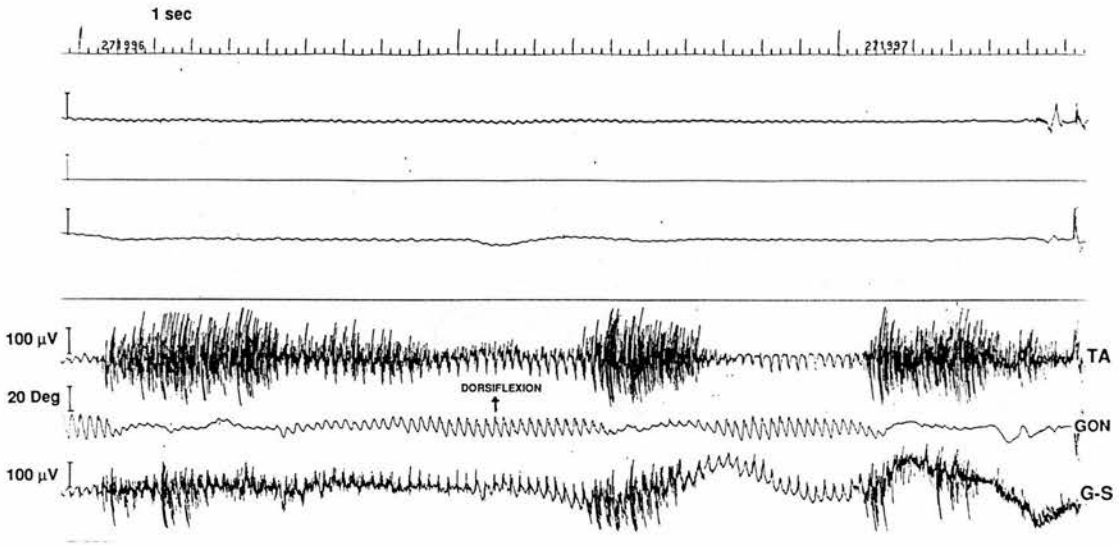
Congenital left hemidystonia in 12 year old girl.

Passive Right ankle dorsiflexion / plantarflexion at 1.5 Hz through a range of 20 degrees.

Silent EMG with movement artefact in Gastrocnemius-Soleus trace. Burst of voluntary activity at end of trace.

TA: Tibialis anterior, G - S Gastrocnemius-Soleus, GON: goniometer. Paper speed 1.5 cm/sec.

Figure 6.3.4 (ii) Unresisted passive alternating movements at the ankle.
Note the restlessness in the 'resting' dystonic limb muscles and bursts of co-contraction of TA and G-S muscles in the non-dystonic limb 3s after the passive stretches cease.



271996/7

Congenital left hemidystonia in 12 year old girl.

Passive Left ankle dorsiflexion / plantarflexion at 3.7 Hz through joint range 14 - 20 degrees.

Three bursts of spontaneous cocontraction abolishes movement at the ankle joint: typical rigid behaviour.

TA: Tibialis anterior, G - S Gastrocnemius-Soleus, GON: goniometer. Paper speed 1.5 cm/sec.

Figure 6.3.3 (iii) Co-contraction abolishes passive motion

6.3. 5 Passive ankle goniometry.

The maximum imposed passive plantarflexion, resting plantarflexion, passive dorsiflexion and passive joint ranges of hemiparetic and nonparetic limbs are shown in table 6.3.1 and typical joint positions in figure 6.2.2a-d. The angles of resting plantarflexion and passive plantarflexion between nonparetic and hemiparetic limbs did not differ statistically. All ankle joints could be passively dorsiflexed to beyond 90° (neutral), the nonparetic more so than hemiparetic limbs ($p < 0.0002$). The overall passive joint ranges were just greater on the nonparetic side by about 8° ($p<0.02$) but the effective joint ranges, ie the angular displacement between the resting angle and maximum passive dorsiflexion, did not differ.

Table 6.3.5 Passive and active hindfoot ankle goniometry and ankle dexterity of non-paretic (NP) and hemiparetic (HP) limbs.

		NP 99% CI		HP 99% CI		
Ankle Position	n	Lower	Upper	Lower	Upper	Paired t Test
Passive PF °	14	48.5	58.3	48.2	59.5	NS
Resting PF °	12	68.5	80.1	68.9	75.3	NS
Passive DF °	14	101.6	108.9	93	100.4	$p < 0.0002$
Passive Range °	14	46.5	57.1	35.5	50	$p < 0.02$
Effective Range °	12	24.2	36.5	18.5	30.3	NS
Active Range °	13	19.2	35.2	2.7	13.6	$p< 0.0003$
Ankle Dexterity Hz	13	1.5	2.8	0.2	1.0	$p< 0.0002$

NP=Nonparetic, HP=Hemiparetic. CI=confidence intervals. n=number of cases.
90°=foot at right angle to tibia.A joint angle of >90° indicates dorsiflexion and a joint angle of <90° indicates plantarflexion beyond neutral. PF=plantarflexion; DF=dorsiflexion,
Range= (Passive DF)-(Passive PF); Effective Range=(Passive DF)-(Resting PF).
Ankle dexterity=frequency of voluntary alternating plantarflexion and dorsiflexion.

6.3.6 Voluntary ankle dexterity (AD) and active range of movement.

Typical examples of the difference in ankle dexterity (AD) between hemiparetic and nonparetic limbs have been described in section 6.3.3 and illustrated in figures 6.3.3 (i-iv) which display the voluntary ankle movements and corresponding voluntary dorsiflexor (TA) and plantarflexor (PF) muscle EMG activity.

Ankle dexterity and amplitude of voluntary motion were greatly reduced for hemiparetic limbs ($p<0.0002$ and $p<0.0003$ respectively, table 6.3.5. In 5/14 cases the

hemiparetic limb had no dexterity at all, i.e. a complete loss of discreet movements at the hemiparetic ankle, and in the remaining 8/14 the mean hemiparetic ankle dexterity was 1.1Hz. The relationship between the measures of dexterity and reflex excitability are examined below.

6.3.7 Reflex excitability.

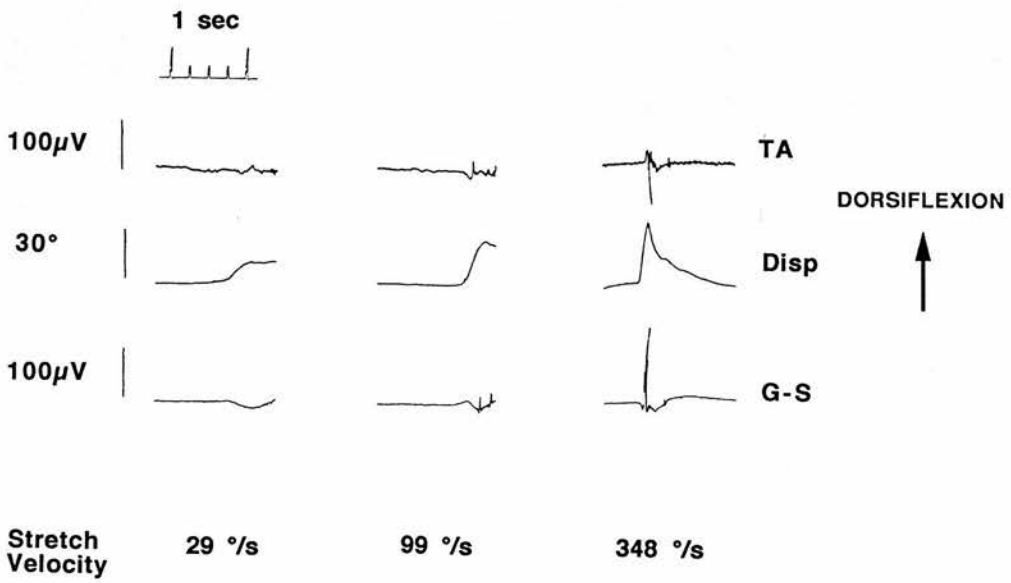
6.3.7.1 Qualitative considerations

Irrespective of the use of ramp or sinusoidal stretches, nonparetic and hemiparetic leg muscles were initially silent till a critical threshold speed of stretch was achieved and this was termed the reflex threshold. A typical example of ramp stretching is given in figure 6.3.7.1 for an 8 year old boy (case 16) with a right hemiparesis. The foot is first passively dorsiflexed at a gentle angular velocity of 29°/s, which is just greater than normal walking speed (Hufschmidt and Mauritz, 1985), but which fails to evoke a stretch reflex. At dorsiflexion velocities of 99°/s, a very weak reflex discharge is apparent, the amplitude of the reflex discharge increasing dramatically as the dorsiflexion angular velocity of stretch increases to 348°/s.

6.3.7.2 Reflex velocity EMG gain.

With increasing speeds of ramp stretch, G-S muscles showed a strong linear increase in reflex EMG and this velocity-dependent increase is referred to as the reflex gain. Group ramp stretch velocity-reflex EMG plots for 14 children are given in figures 6.3.7.2: the top scatter plot gives the EMG gain in raw EMG microvolts (μV) for nonparetic and hemiparetic limbs. If the EMG gain is normalised to the maximum value for each leg, in each child (fig. 6.3.7.2 bottom plot) the nonparetic and hemiparetic EMG gain is largely similar. Figure 6.3.7.3 plots the normalised velocity-EMG gain separately for the nonparetic (top) and hemiparetic (bottom) limbs. For two subjects, the hemiparetic reflex discharges appear to *saturate* at very high angular velocities of stretch, up to about 800°/s.

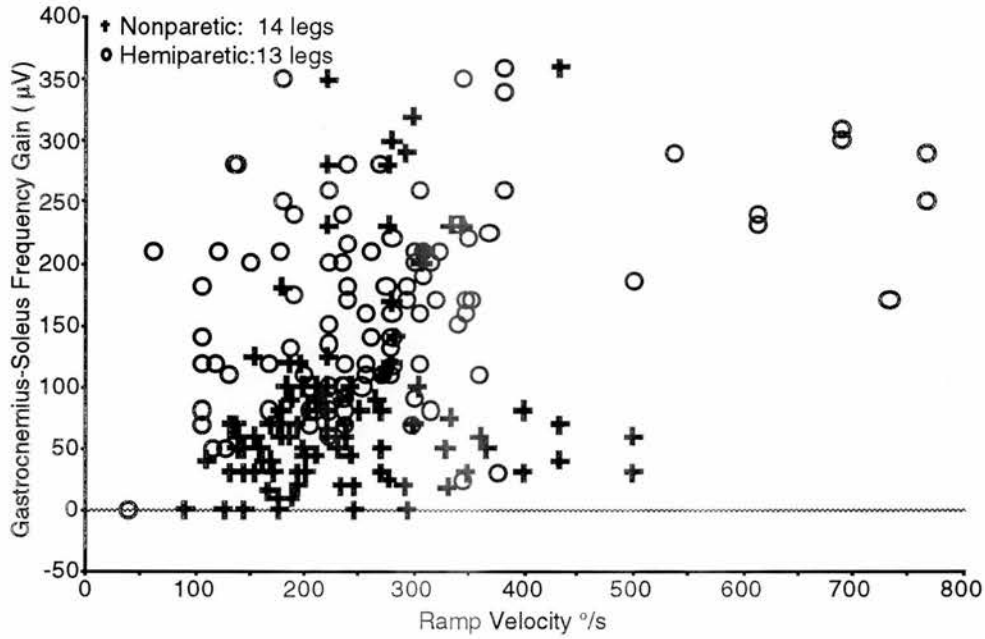
The individual ramp stretch velocity-reflex EMG plots for 14 children are given in figures 6.3.7.4(i-iv). It should be noted that in case 10, no nonparetic reflex velocity threshold data was obtained.



Ramp stretch of hemiparetic TA and G-S muscles with increasing velocity. 8 year old boy with right hemparesis 2° to prematurity.

TA = Tibialis Anterior
G-S = Gastrocnemius-Soleus
Disp = Ankle displacement

Figure 6.3.7.1 Ramp stretching and reflex velocity gain



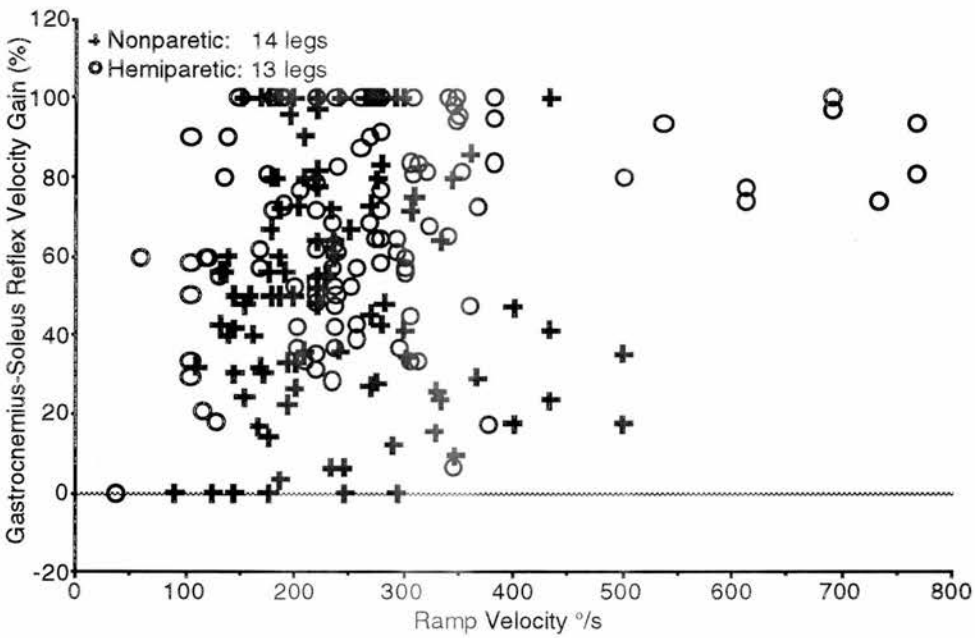


Figure 6.3.7.2 Raw (top) and normalised(bottom) reflex velocity EMG gain.

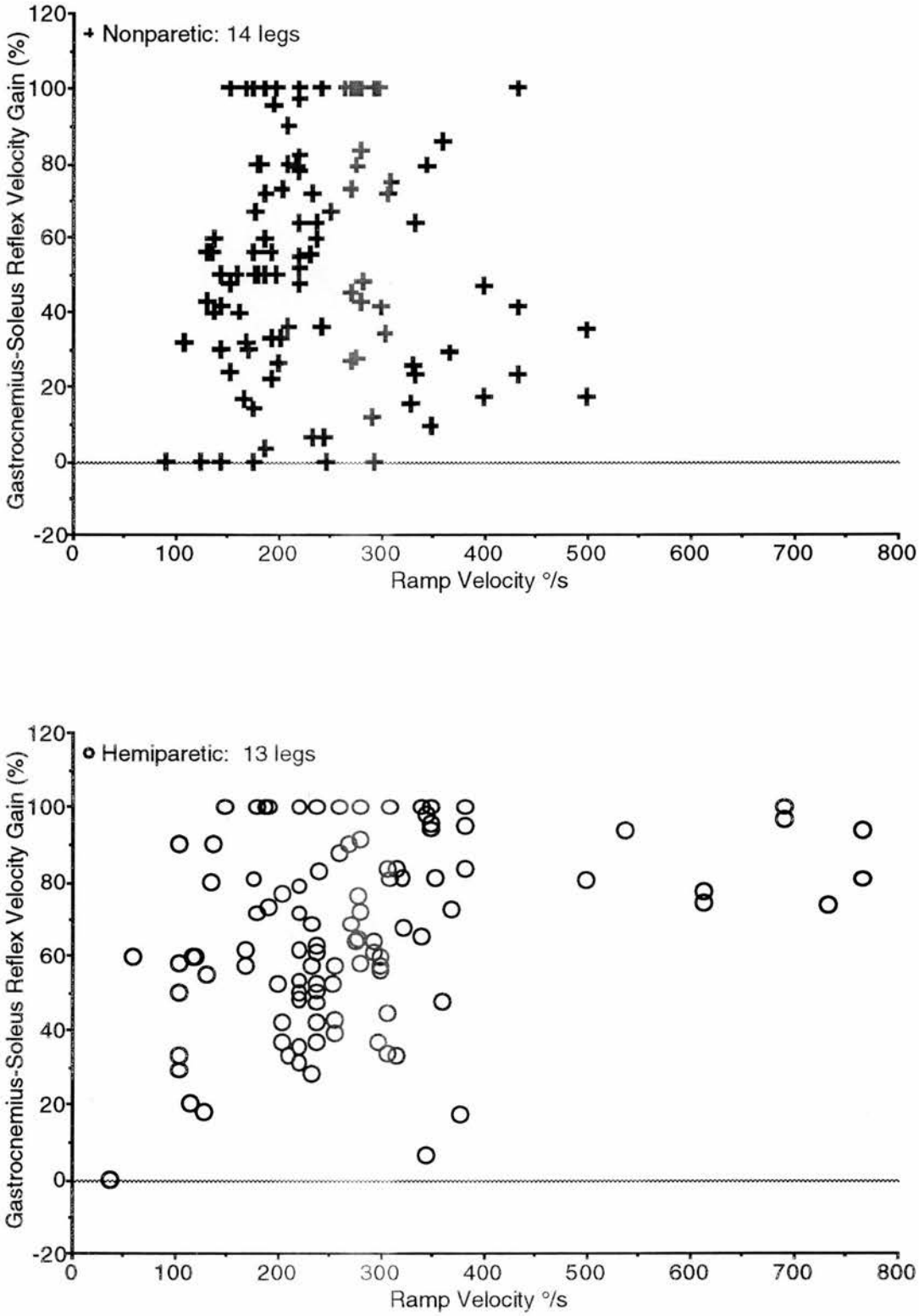


Figure 6.3.7.3 Separate nonparetic (top) and hemiparetic (bottom) reflex EMG gain.

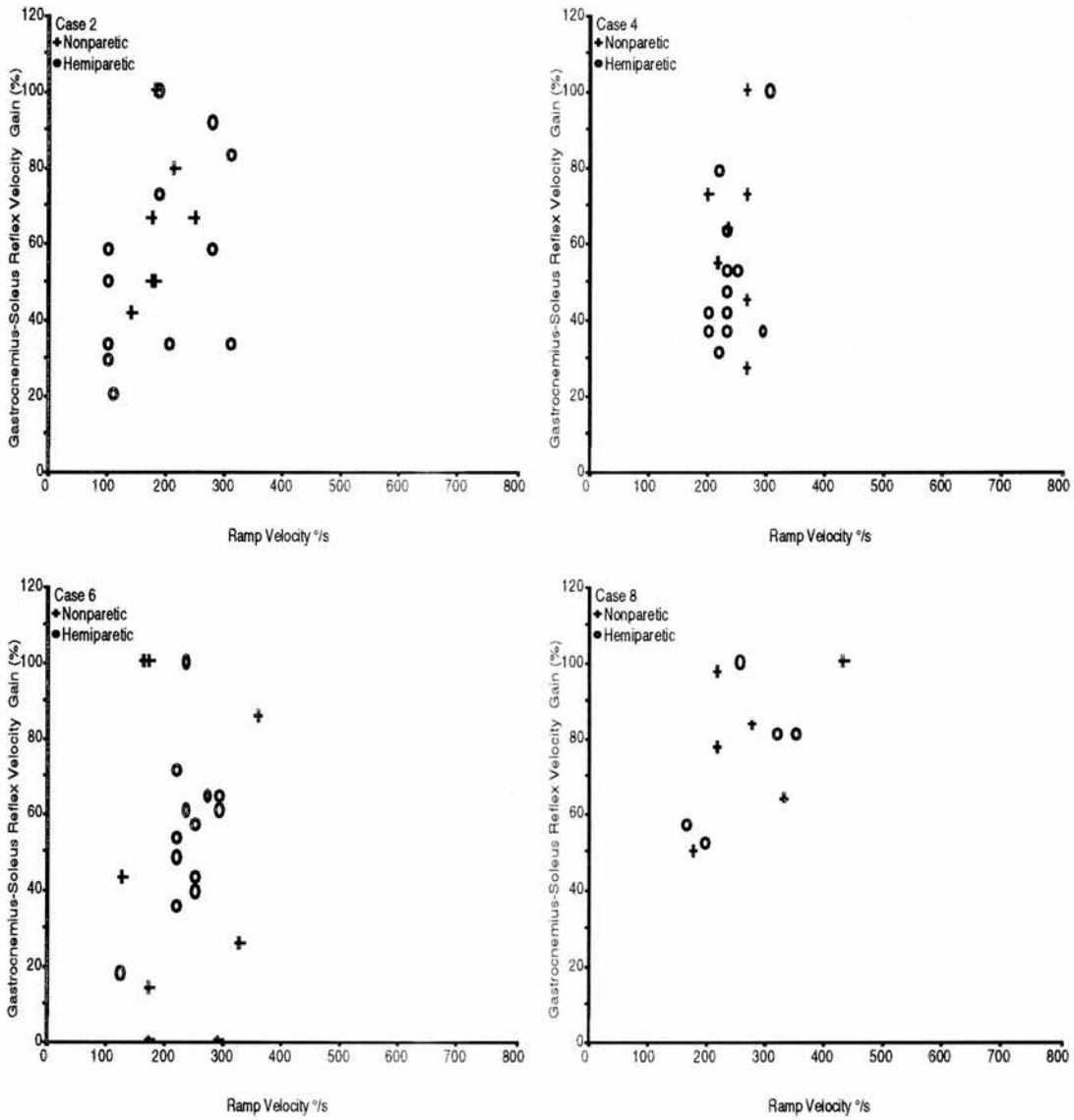


Figure 6.3.7.4.(i) Reflex velocity threshold and gains: Cases 2,4,6 and 8.
For explanation, see text.

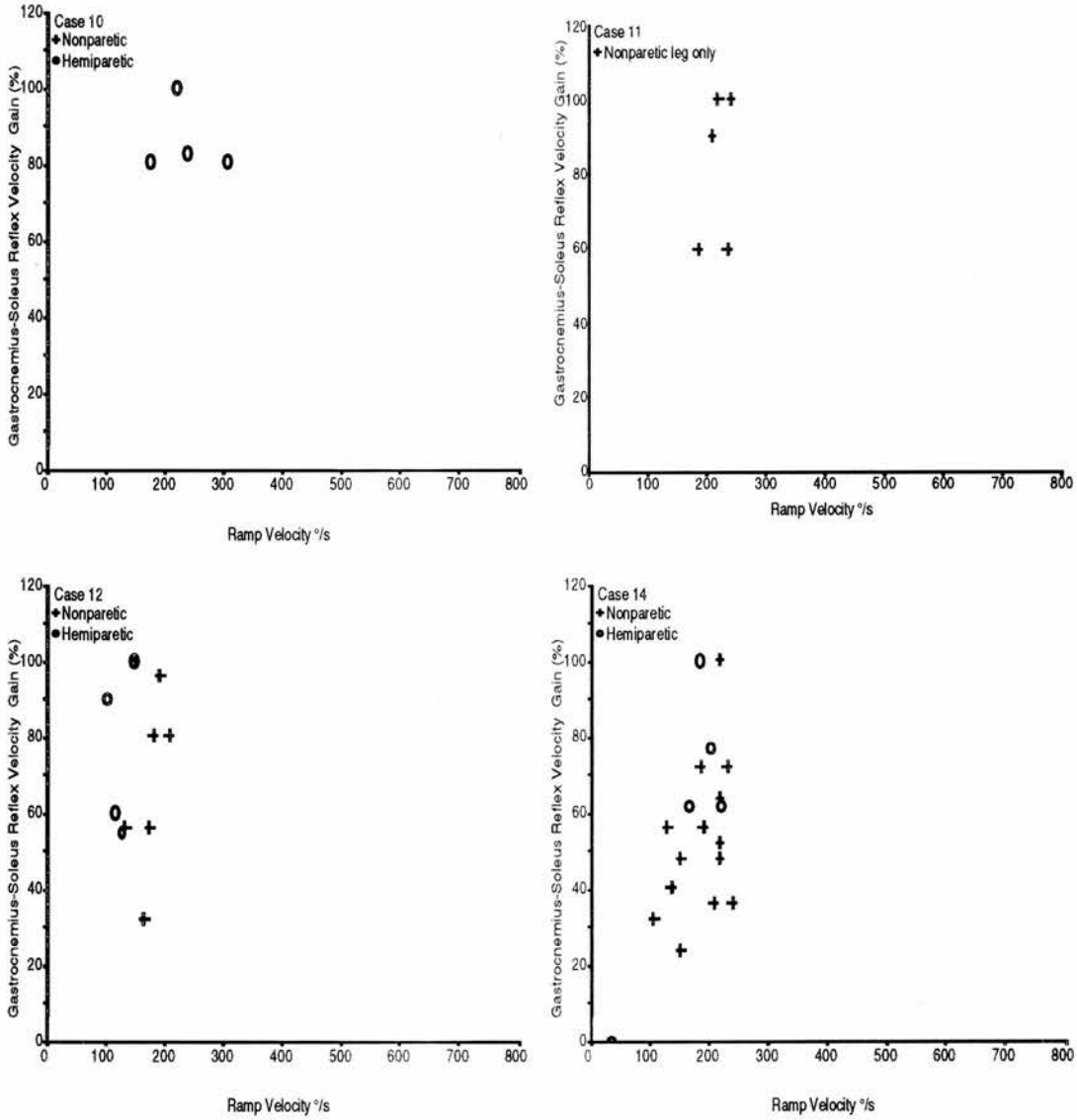


Figure 6.3.7.4 (ii) Reflex velocity threshold and gains: Cases 10, 11,12 and 14.
For explanation, see text.

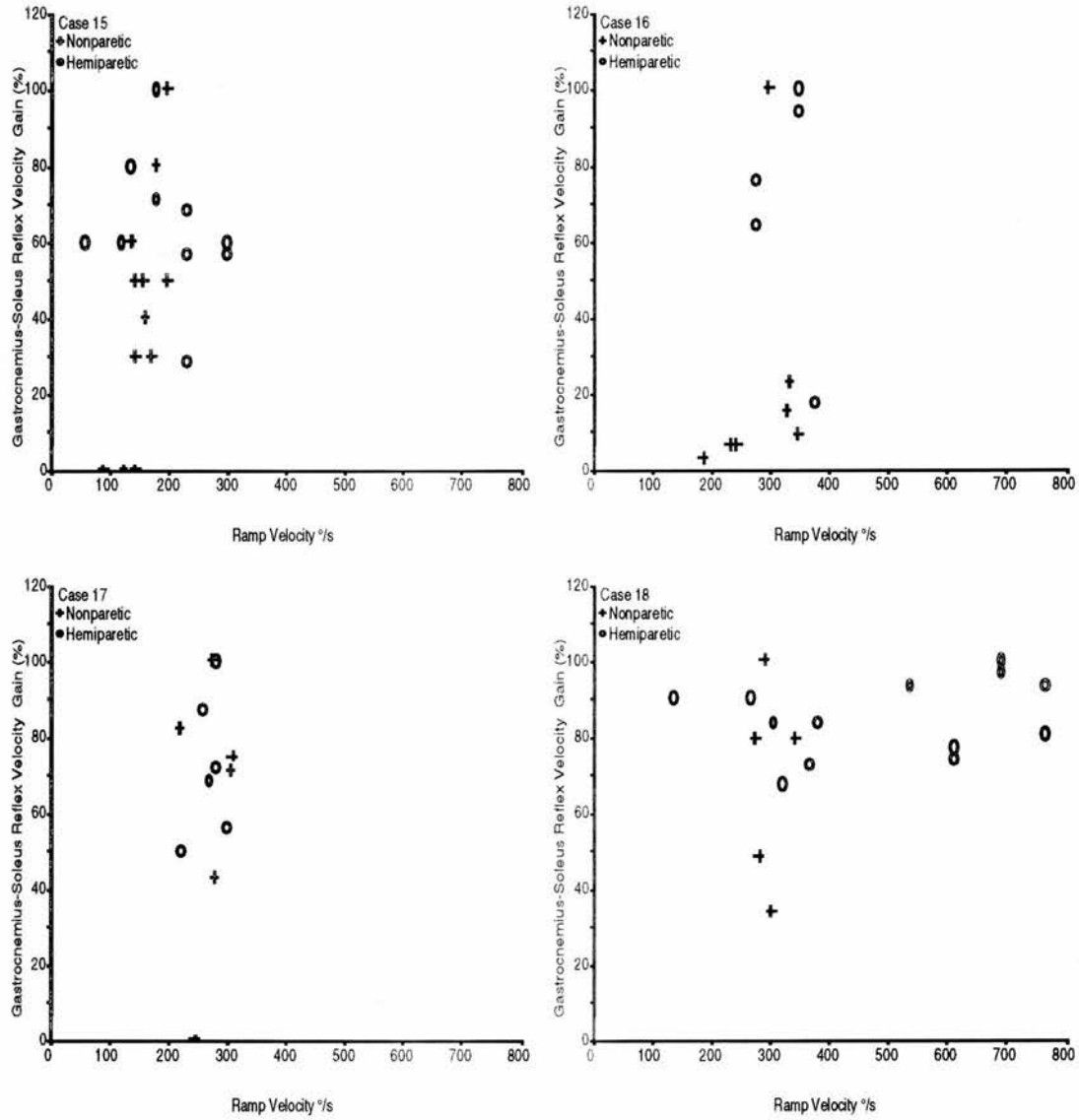


Figure 6.3.7.4 (iii) Reflex velocity threshold and gains: Cases 15,16, 17 and 18.
For explanation, see text.

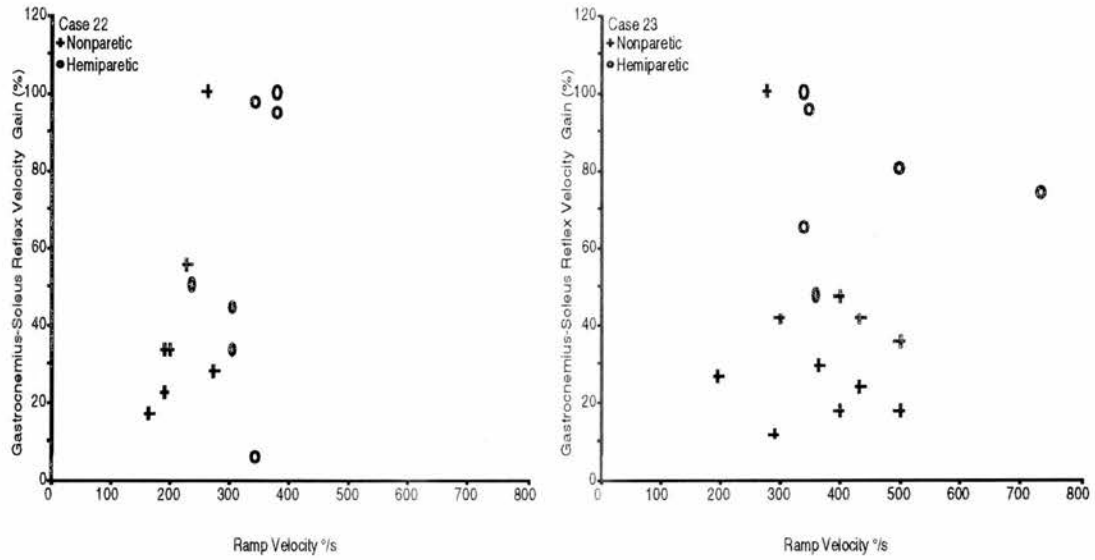


Figure 6.3.7.4 (iv) Reflex velocity threshold and gains: Cases 22 and 23.
For explanation, see text.

The overall behaviour of the reflex -velocity plots is very similar, in general exhibiting maximum reflex excitability at maximum velocities of stretch. In few cases was the actual reflex velocity threshold measured since the 'take off point" or onset of the EMG discharges, occurred quite abruptly at a critical velocity.

The means, standard deviations, 99% confidence intervals and analysis of variation (ANOVA Statview +Graphics) for the nonparetic and hemiparetic ramp velocity stretches respectively are given in table 6.3.7.2. For the group as a whole, there was significant difference in ramp velocity of stretch between nonparetic and hemiparetic limbs, but this is undoubtedly due to cases 18 and 23 in whom stretch velocities for the hemiparetic limbs exceeded 500°/s. If the analysis excludes rates of stretch greater than 550°/s, there is no significant difference between the nonparetic and hemiparetic stretch velocities.

The raw reflex velocity EMG gain for hemiparetic limbs (158μV +/- 76) is almost twice as great as that of the nonparetic limbs (84.8μV +/-80.6), $p<0.0002$. However, if the normalised reflex velocity gain is compared, the differences between means are still significant ($p<0.0016$) but appear less dramatic, being 56.6%+/-31.5 and 64.4+/-24.7 for nonparetic and hemiparetic limbs respectively.

Table 6.3.7.2 Gastrocnemius-Soleus reflex gain with ramp velocity stretches

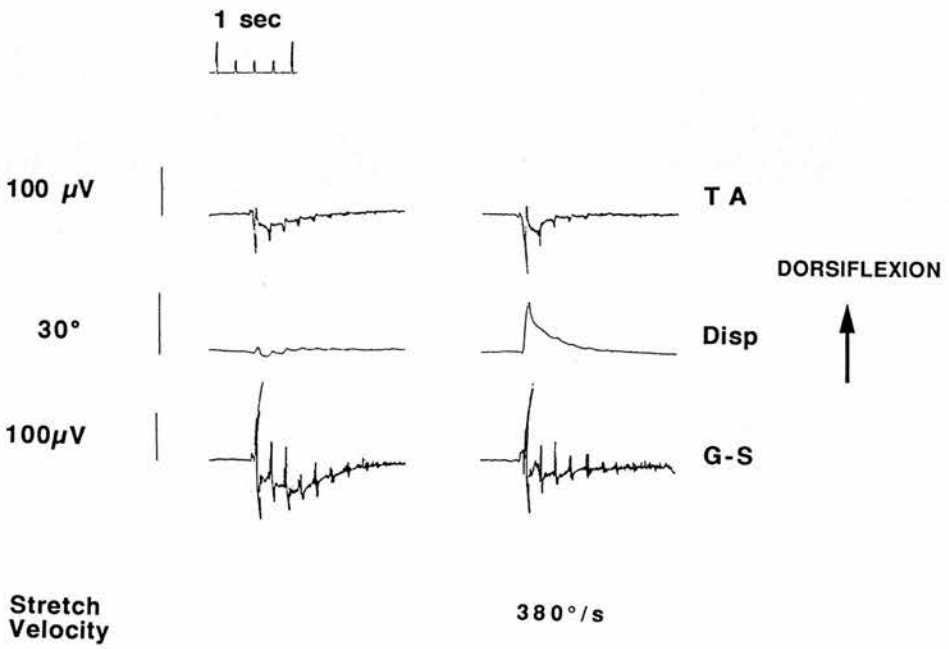
	Nonparetic	**Limb**			**Hemiparetic**	**Limb**			
Variable	mean	SD	99% CI lower	CI upper	mean	SD	99% CI lower	CI upper	ANOVA
Ramp Velocity (°/s)	**234.0**	83.0	212.7	255.3	**245.2**	88.6	222.1	268.2	p=0.35
Raw G-S EMG (μV)	**84.8**	80.6	64.1	105.4	**158.0**	76.0	138.2	177.8	p<0.0002
Normalised G-S EMG (%)	**51.6**	31.5	43.5	59.7	**64.4**	24.7	57.9	70.8	p<0.0016
Clonus Beats (N°)	**1.3**	.99	1.1	1.6	**3.6**	2.6	2.9	4.3	p<0.0002

6.3.8 Clonus beats following ramp stretches.

A typical example of unsustained clonus elicited by manual ramp stretches is given in figure 6.3.8.1 (above). There is no obvious relationship between the velocity of ramp stretch and the frequency of clonus elicited. However the relationship between clonus and joint angle is explored in detail in a later section.

For the group as a whole, the difference in the number of elicited clonus beats (table 6.3.7.2) was twice as great in the hemiparetic limbs, being 1.37+/-0.99 for nonparetic and 3.64+/-2.67 for hemiparetic limbs respectively. But in clinical terms, only a few hemiparetic limbs exhibited greater than five clonus beats (figure 6.3.8.2) in response to manual stretches. No child demonstrated clonus during foot-contact in the gait cycle.

It was difficult from this study to fully understand the relationship between clonus and velocity-dependence. Figure 6.3.8.1 shows that the interval between each clonus discharge is about 200-250ms, which is an extremely long interval: ie about 5-7 times the time for the monosynaptic reflex arc from the G-S muscles in an adult. This latency may be related to intraspinal events, but it seems more likely to depend on the muscle twitch-time which entails a time course of this order of magnitude. Nor is there any clue to the clear decrement in clonus beats evident in figure 6.3.8.1. The present study was unable to shed any further light on the manifestations of clonus, although in a subsequent section which studies the relationship between monosynaptic ankle reflexes and muscle twitch characteristics, evidence for the role of the muscle twitch phenotype in the pathophysiology of clonus is advanced. Aside from the phenomenon of clonus, more graded responses were obtained using sinusoidal stretches, which were considered to reflect a more common pattern of physical perturbation.



Clonus of hemiparetic TA and G-S muscles is independent of stretch velocity. 12 year old boy with left hemiparesis 2° to IUGR.

TA = Tibialis Anterior
G-S = Gastrocnemius-Soleus
Disp = Ankle displacement

Figure 6.3.8.1 Clonus beats and the velocity of manual ramp stretch
There is no apparent relationship between the velocity of stretch and the number of clonus beats evoked.

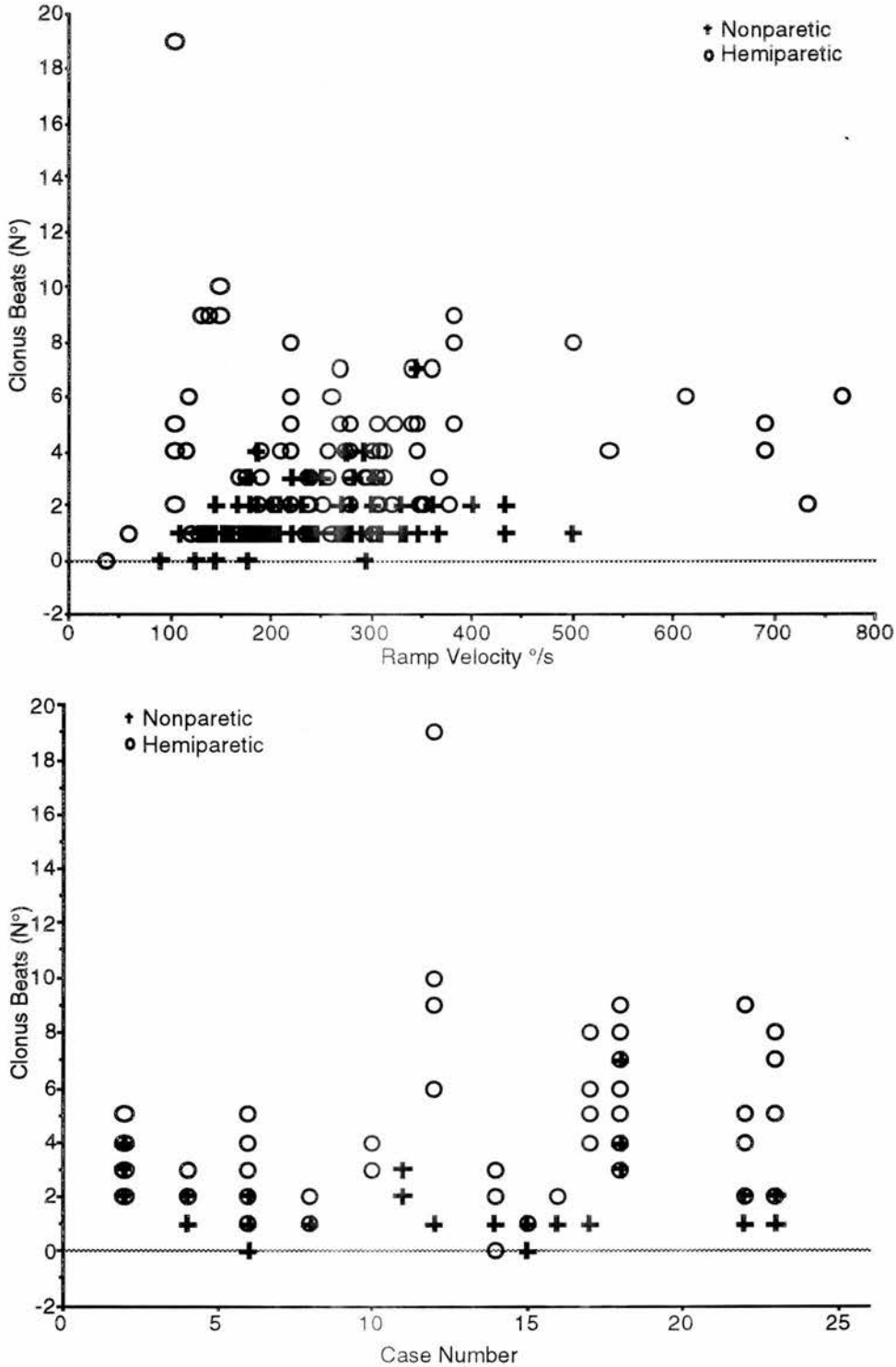


Figure 6.3.8 2 Clonus beats in nonparetic and hemiparetic limbs
Top: Clonus and velocity of ramp stretch. Bottom: Clonus beats according to each case.

6.3.9 Sinusoidal Stretch.

The reflex gastrocnemius-soleus (G-S) EMG against frequency of sinusoidal stretch is shown in figure 6.3.9.1(top) and the corresponding graphical plot of the EMG amplitude against frequency of stretch in figure 6.3.9.1 (bottom) for the hemiparetic limb of case 16, an 8 year old boy with a congenital right hemiplegia. The first EMG reflex discharge begins at just over 1 Hz and increases linearly with faster frequencies of stretch. The intercept of the linear regression equation gives a derived reflex frequency threshold of 1.2 Hz. The slope of the the regression line is 40.3, which is a measure of the reflex frequency gain in % μV/ Hz.

Other examples of sinusoidal stretch are given in figure 6.3.9.2 for case 17, a 6 year-old boy with a congenital right hemiplegia for whom sinusoidal stretches at frequencies of 0.75-3Hz were manually applied to the G-S muscles: in this case the reflex EMG discharges occur between 2-3Hz on the hemiparetic side, but the nonparetic limb is refractory to stretch.

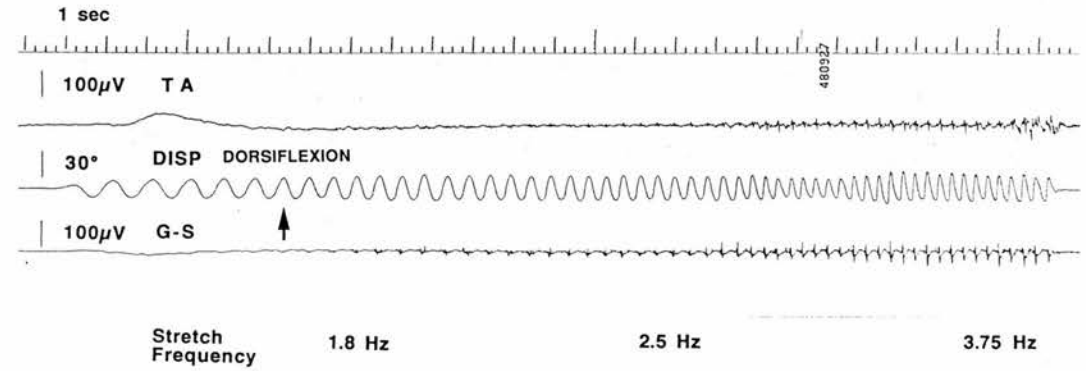
In another example of sinusoidal stretch at the ankle, case 18, with a congenital left hemiparesis , shows evidence of a tonic reflex EMG discharge at low frequencies of stretch: these discharges being in phase with maximum dorsiflexion. As the frequency of passive stretch increases, so the reflex discharge becomes progressively phasic, ie maximal at the midpoint of the joint range, in phase with maximal stretch velocity.

It was easier to apply a graded stretch stimulus using sinusoidal stretches than when employing manual ramps. The results for the 14 children as a group are given in figures 6.3.9.3, and for nonparetic and hemiparetic limbs separately in figure 6.3.9.4 and individual cases in figure 6.3.9.5 (i-iv).

In figure 6.3.9.5, the hemiparetic data appears to rise reasonably steeply with increasing frequencies of stretch between 1-5Hz and then to saturate at sinusoidal frequencies of greater than 5Hz. The rise in reflex EMG is borne out by the individual plots in figures 6.3.9.6 (i-iv), which also show the saturation phenomenon most clearly for individual cases 2, 10, 18 and 22.

No reflex frequency threshold was reached in 8 nonparetic limbs in cases 2,4,6,8,11,12, 17 and 23, despite adequate frequencies of stretch (fig. 6.3.9.6 i-iv:arrows).

Table 6.3.9.1 summarises group data with respect to means, standard deviations, 99% upper and lower confidence intervals(CI) and analysis of variation (ANOVA) between nonparetic and hemiparetic limbs for sinusoidal frequency, raw and normalised reflex EMG.



Sinusoidal stretch of hemiparetic TA and G-S muscles at increasing frequency. 8 year old boy with right hemiparesis 2° to prematurity.

TA = Tibialis Anterior
G-S = Gastrocnemius-Soleus
Disp = Ankle displacement

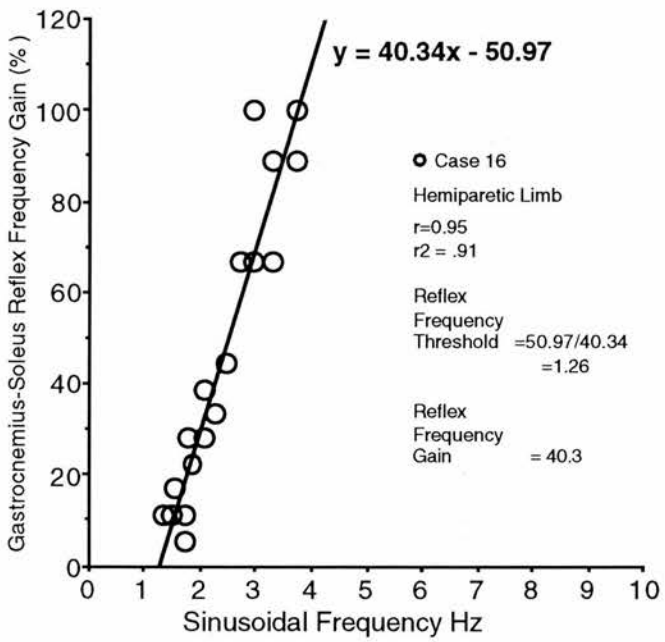
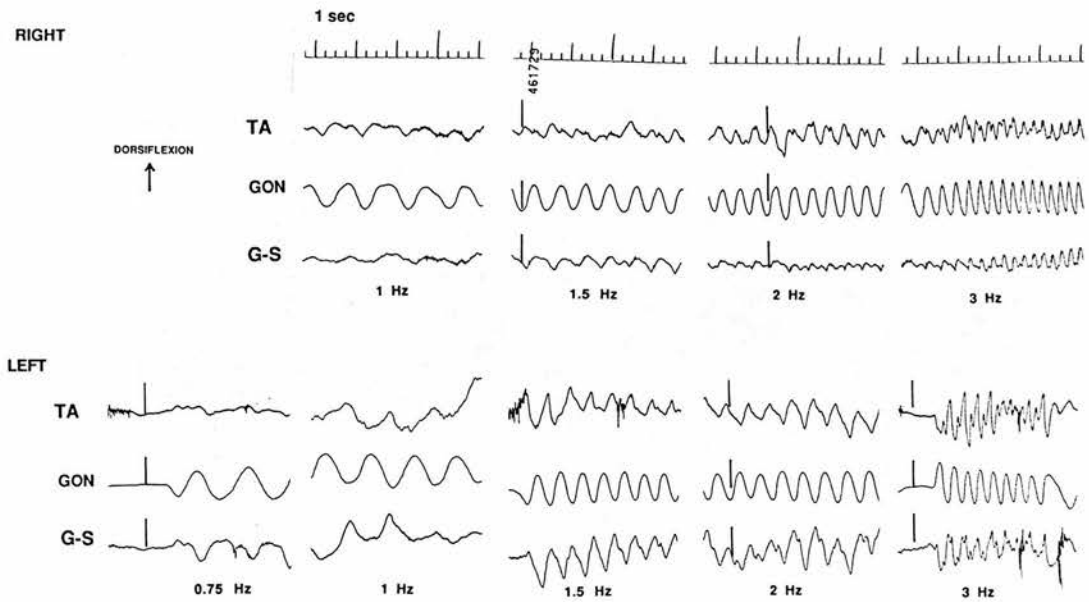


Figure 6.3.9.1 Sinusoidal stretch and the hemiparetic G-S reflex EMG threshold and gain. There is a clear linear increase in reflex G-S muscle discharges with frequency of stretch (top). The reflex EMG threshold can be derived from the x-intercept and the reflex EMG gain is determined by the slope of the linear regression equation (bottom):y=40.34X - 50.97 while the Reflex Frequency Gain (RFG)= 40.3%per Herz of sinusoidal stretch frequency.



Passive sinusoidal movement of the ankles, varying amplitude, frequency and velocity.

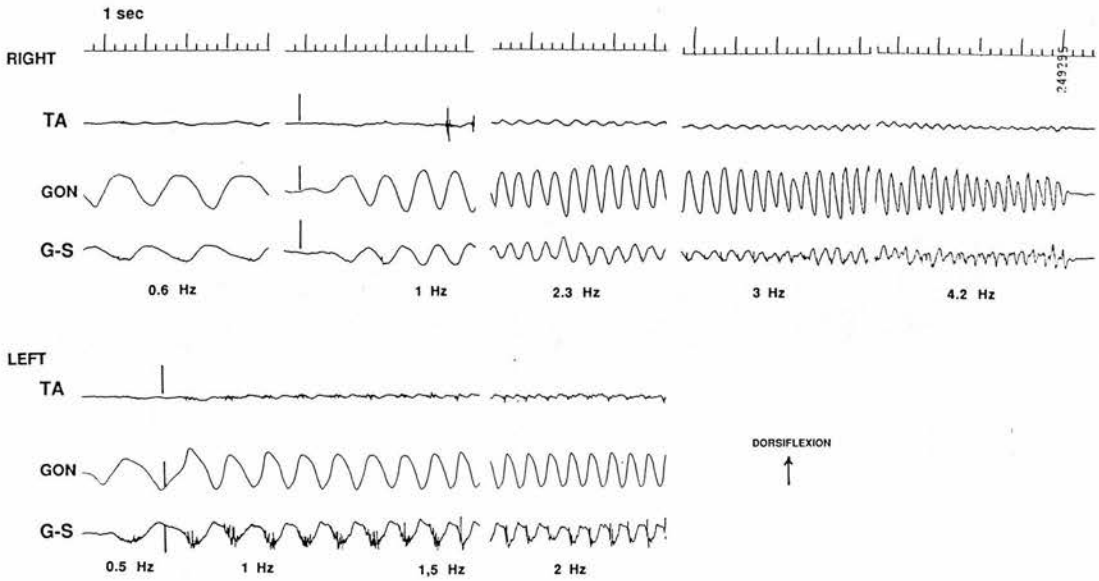
Case 17. Congenital Right Hemiplegia. Age 6.5 years.
Clinical evidence of "tonic calf spasticity", markedly reduced ankle compliance and equinus gait. Patient prone, "at rest".

	Amplitude	Frequency	EMG
Right (affected) ankle	20°	1, 1.5, 2, 3 Hz	silent
Left (normal) ankle	20°-27°	0.75, 1, 1.5, 2, 3 Hz	silent

Sinusoidal stretching elicits no reflexes in the affected or "normal" TA and G-S muscles.

TA: Tibialis Anterior EMG, G-S: Gastrocnemius-sSoleus EMG, GON: Goniometer.
Calibration bars: EMG 100μV, goniometer 20°, time 1 sec.

Figure 6.3.9.2. Bilateral sinusoidal stretching



Passive sinusoidal movement of the ankles, varying amplitude, frequency and velocity.

Case 18. Congenital Left Hemiplegia. Age 12 years.
Clinical evidence of "tonic calf spasticity", mildly reduced ankle compliance but with some heel contact during gait cycle. Patient prone, "at rest".

	Amplitude	Frequency	EMG
Right (normal) ankle	20°-40°	0.6, 1, 2.3, 3, 4.2 Hz.	silent
Left (affected) ankle	30°	0.5, 1, 1.5, 2 Hz.	? tonic

Tonic EMG co-contraction in Left (affected) TA and G-S most pronounced at low frequencies and in phase with displacement i.e. muscle length, not velocity. This muscle contraction contributes to the perceived resistance to passive stretch. Is this a loading /unloading response on actively contracting muscles as suggested by the lack of velocity-dependence?

TA: Tibialis Anterior EMG, G-S: Gastrocnemius-Soleus EMG, GON: Goniometer.
Calibration bars: EMG 100μV, goniometer 20°, time 1 sec.

Figure 6.3.9.3 Another example of passive sinusoidal stretching.
At cycles of stretch of 2Hz, the earlier tonic G-S stretch reflex , in phase with length is now clearly phasic, ie in phase with velocity.

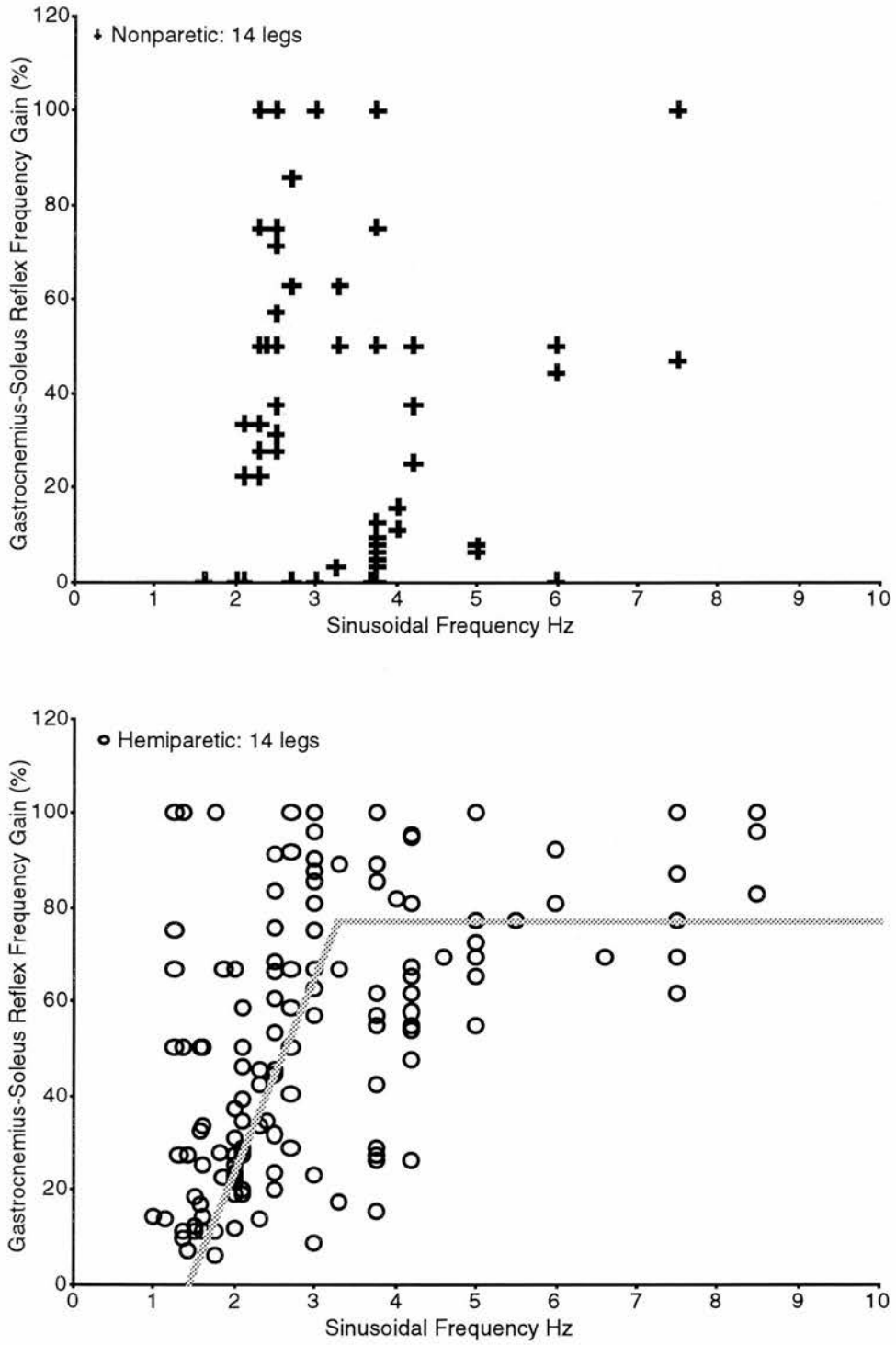


Figure 6 3.9.5 Nonparetic and hemiparetic reflex frequency discharges.
Top: The nonparetic limbs failed to reach the reflex frequency threshold in 8/14 cases despite high frequencies of stretch (see cases 2,4,6,8, 11,12, 17, and 23 below).
Bottom: For the hemiparetic limbs, the reflex discharges appear to rise steeply between 1-5Hz and then to 'saturate' after 5Hz. This suggests a sigmoidal relationship (dotted line). See also individual subject plots below.

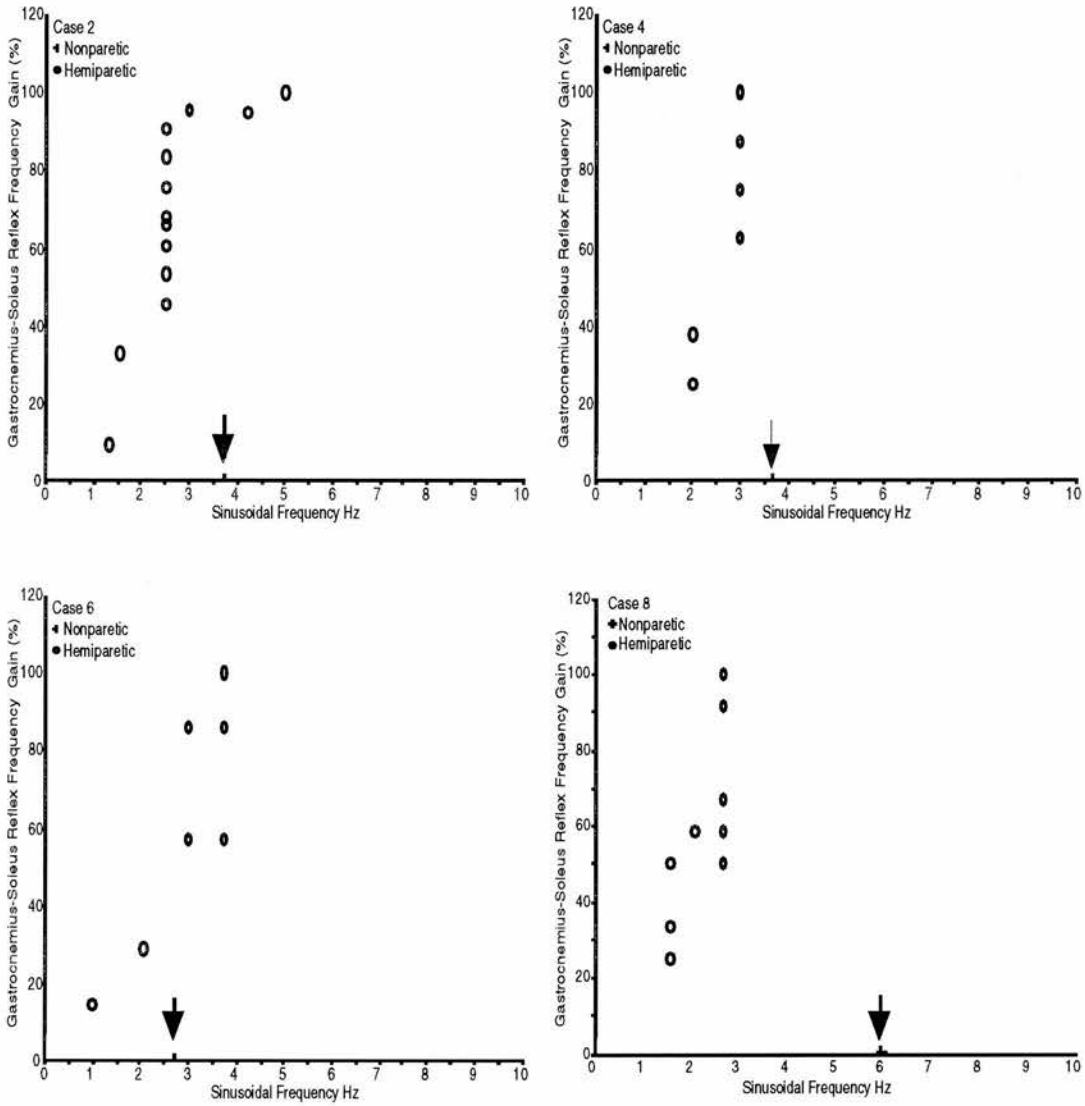


Figure 6.3.9.6 (i) Reflex frequency G-S EMG discharges: cases 2, 4, 6 and 8.
Note absence of the nonparetic sinusoidal stretch reflex threshold, despite high stretch frequencies (arrows). Saturation of the hemiparetic stretch reflex gain is seen in case 2.

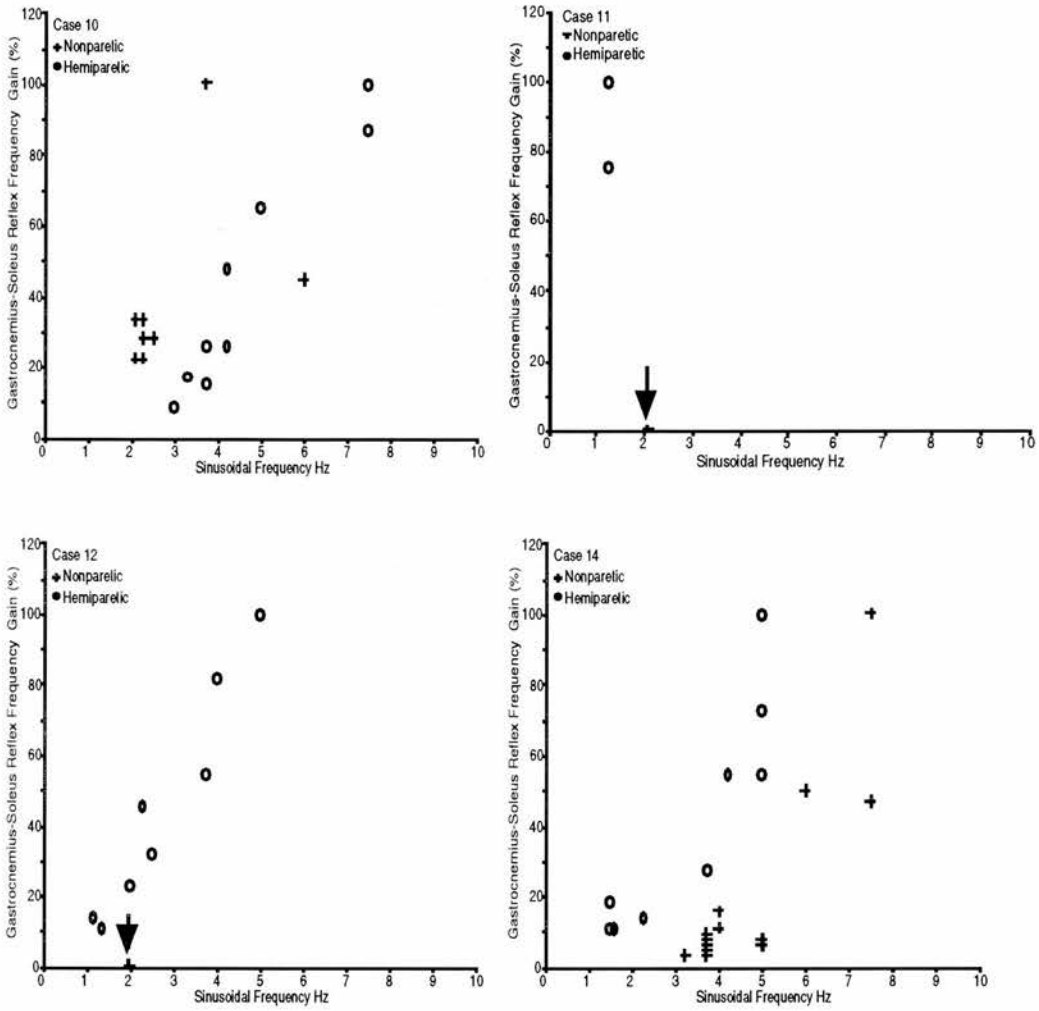


Figure 6.3.9.6 (ii) Reflex frequency G-S EMG discharges: cases 10, 11, 12 and 14.
High frequencies of stretch fail to elicit the stretch reflex in the nonparetic limbs of cases 11 and 12.(arrows).In case 10, the reflex frequency EMG discharges occur at lower frequencies in the nonparetic limb but in case 14, the nonparetic limb has a higher reflex threshold. Saturation of the hemiparetic stretch reflex gain is seen in case 10.

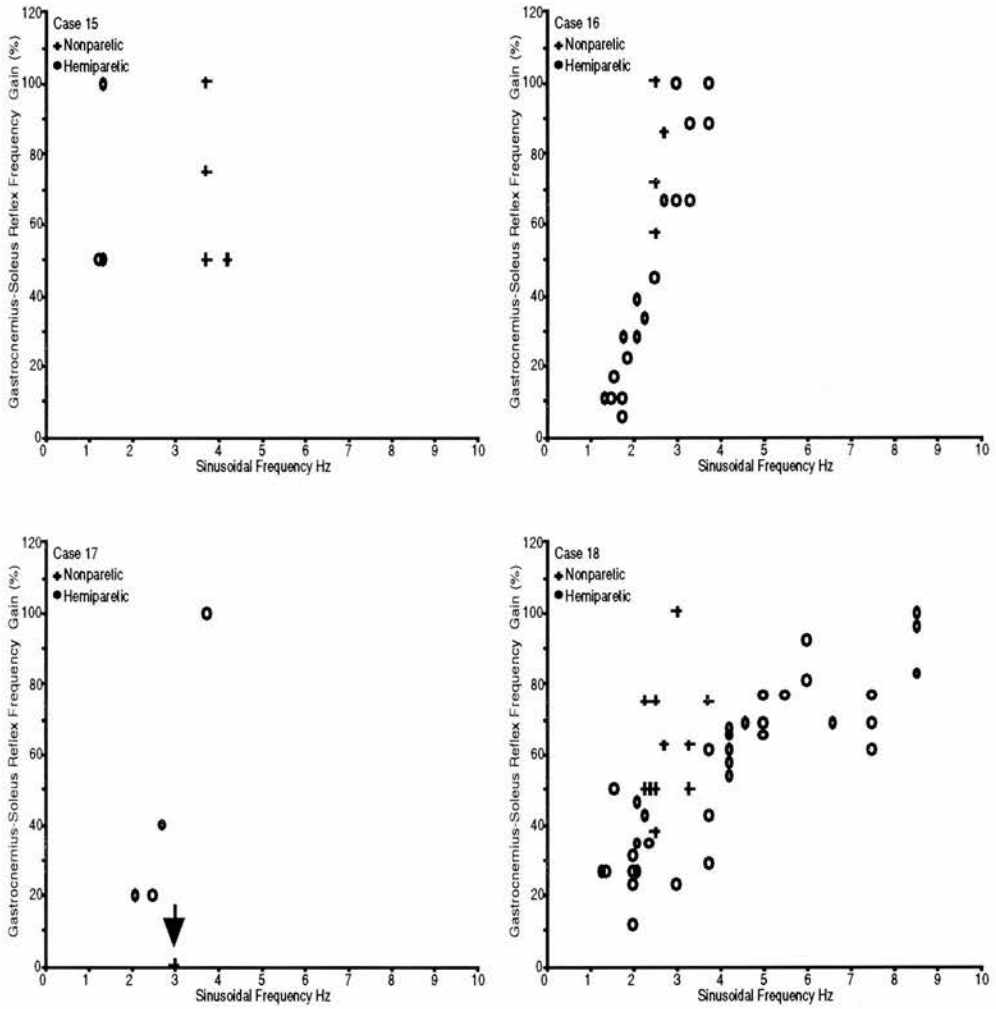


Figure 6.3.9.6 (iii) Reflex frequency G-S EMG discharges: cases 15, 16, 17 and 18.
There is a failure to reach the nonparetic reflex frequency threshold in case 17: arrowed. Saturation of the hemiparetic stretch reflex gain is seen in case 18.

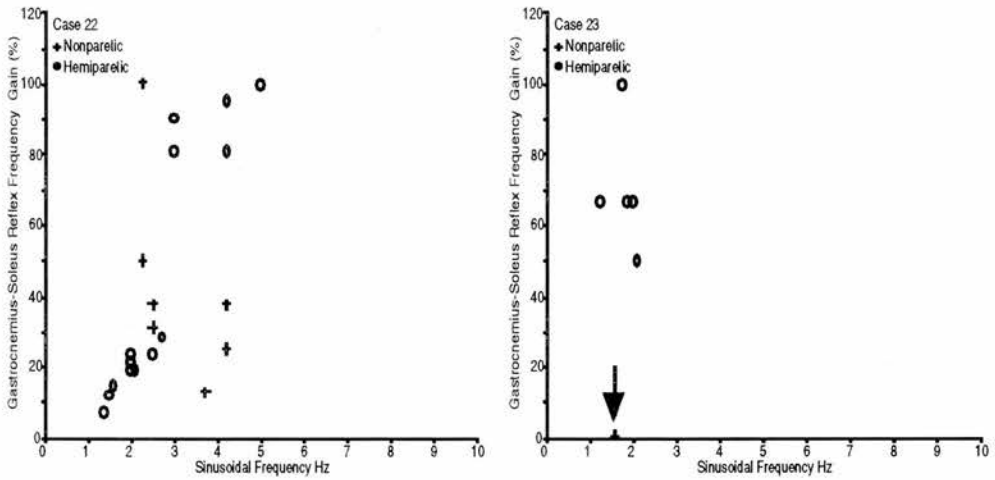


Figure 6.3.9.6 (iv) Reflex frequency G-S EMG discharges: cases 22 and 23.
Case 22: a possible sigmoidal pattern for the hemiparetic limb. Case 23 with a dystonic left hemisyndrome: no reflex frequency threshold is reached on the nonparetic side (arrowed).

There was no difference in sinusoidal stretch frequency between nonparetic (3.4+/-1.2 Hz) hemiparetic (3.0+/-1.6Hz) limbs.

The raw sinusoidal G-S reflex EMG gain was twice as high in the hemiparetic (70.3+/-62.8μV) as compared with the nonparetic reflex gain (35.5+/-46.4μV), which was highly statistically significant, $p<0.0002$.

When normalised, the G-S reflex frequency EMG difference appears less marked: nonparetic EMG 43.4+/-33.1% and hemiparetic EMG 55.3+/29.2%, $p<0.006$. This is undoubtedly due to the failure to reach a reflex frequency threshold in six of the nonparetic limbs. For the remainder

Table 6.3.9.1 Gastrocnemius-Soleus reflex frequency gain with sinusoidal stretches

n=14	**Nonparetic Limb**				**Hemiparetic Limb**				
Variable	mean	SD	99% CI lower	CI upper	mean	SD	99% CI lower	CI upper	ANOVA
Sinusoidal Stretch frequency (Hz)	3.4	1.2	3.0	3.8	3.0	1.65	2.7	3.3	p=0.1366
Raw Reflex EMG (μV)	35.5	46.4	20.6	50.5	70.3	62.8	58.4	82.1	p<0.0002
Normalised Reflex EMG (%)	43.4	33.1	32.7	54.0	55.3	29.2	49.8	60.8	p<0.006

Table 6.3.9.2 Gastrocnemius-Soleus reflex frequency gain with sinusoidal stretches

n=6	**Nonparetic**		**Limb**		**Hemiparetic**		**Limb**		
Variable	mean	SD	99% CI lower	CI upper	mean	SD	99% CI lower	CI upper	ANOVA
Sinusoidal Stretch frequency (Hz)	3.44	1.23	3.0	3.8	3.42	1.97	2.9	3.9	p=0.989
Raw Reflex EMG (μV)	40.3	47.6	24.0	56.6	91.5	73.3	73.2	109.7	p<0.0002
Normalised Reflex EMG (%)	49.2	30.9	38.5	59.8	50.1	29.9	42.7	57.6	p=0.712

Excludes cases 2,4,6,8,11,12, 17 and 23 in which no nonparetic reflex frequency threshold was reached.

By excluding the eight cases in whom no reflex frequency threshold was reached in the nonparetic limbs, table 6.3.9.2 indicates that the remaining limbs show no differences in reflex frequency gain for normalised G-S EMG amplitudes: the mean nonparetic reflex EMG being 49.2+/-30.9% compared with a hemiparetic reflex EMG of 50.1+/-29.9%.

6.4 Summary of results.

6.4.1 Patterns of EMG activation.

i. Unless there is co-existing dystonia, the resting EMG in hemiparetic limbs is silent.

ii. The EMG of the hemiparetic limb exhibits less activity in standing than the nonparetic limb, and this includes hemidystonic lower limbs too.

iii. The EMG amplitude during isometric plantarflexion and dorsiflexion is always reduced on the hemiparetic side, indicating a predominance of small motor units and /or the inaccessability to recruitment of larger motor units.

iv. During isotonic tasks, such as alternating movements at the ankle, nonparetic limbs were able to modulate the frequency and amplitude of motion with corresponding modulation of motor unit recruitment and the presence of a triphasic EMG discharge pattern between agonist and antagonist muscles.

v. Hemiparetic muscles displayed low-amplitude, poorly differentiated EMG activity during attempted alternating movements at the ankles and toes, associated with low

amplitude co-contraction.

vi. The single hemidystonic limb exhibited frequent bursts of high amplitude co-contraction, both at rest and especially during attempted voluntary movements.

vii. Passive movements could be disturbed by voluntary agonist 'assistance in which the assisting EMG activity was 180° in phase advance of the intended peak motion.

viii Voluntary and involuntary resistance during passive lengthening, resulted in tonic EMG discharges that were, by definition in phase with maximum angular displacement, followed by a 'muscle silent' period which occurred during passive shortening.

ix Velocity-dependent reflex discharges occurred, by definition, in phase with maximum angular velocity and 90° in phase advance of maximum passive angular displacement.

x The timing of the effect of such a reflex discharge would occur with a 180° phase lag on the reflex EMG discharge.

6.4.2 Active joint range and ankle dexterity.

i. The active joint range was greatly reduced on the hemiparetic side, despite equivalent passive joint ranges between the sides

ii. Ankle dexterity (AD) frequencies for nonparetic and hemiparetic sides, defined as the maximum frequency of voluntary alternating plantarflexion / dorsiflexion, were significantly lower for hemiparetic(HP) ankle compared to nonparetic (NP) limbs: mean HPAD=0.6 (99%CI:0.2-1) HZ and mean NPAD=2.1Hz (99%CI: 1.5-2.8) HZ

6.4.3 Ramp stretches.

i. For equivalent ramp velocity stretches, no reflexes were elicited at normal walking angular velocities of stretch in any limb (ie velocities<29°/s).

ii. There appeared to be significant differences in Gastrocnemius-Soleus muscle raw and normalised reflex velocity EMG gain between nonparetic and hemiparetic limbs, the latter being more excitable.

iii. No clear relationship between clonus and stretch velocity could be established.

iv. Hemiparetic limbs exhibited more than 5 beats of clonus in only 4/14 cases.

6.4.4 Sinusoidal stretches.

i. Sinusoidal stretches allowed a more graded reflex response to be measured.

ii. For the population as a group, the normalised hemiparetic reflex frequency EMG amplitude, rose steeply between 1 -5Hz and then appeared to saturate with faster stretch frequencies. This was borne out by the individual frequency-EMG plots in four cases.

ii. For sinusoidal stretches, the major difference between the nonparetic and hemiparetic limbs seems to be that in a significant proportion(8/14), the nonparetic limbs are inexcitable.

iii. If the 8/14 refractory cases are excluded from the analysis, the remaining 6/14 cases show no differences in normalised reflex frequency EMG gain at equivalent sinusoidal frequencies.

7. Soleus Muscle Reflex Excitability and the Joint Angle.

7.1 Background.

It has been known for more than 40 years that changes in the joint angle alter reflex excitability (Magladery et al 1951, Paillard 1959). since then, the mechanisms underlying the joint angle modulation and its significance for the physiology and pathophysiology of motor function have become clearer.

Herman (1969, 1970) indicated that the joint angle corresponding to maximum excitability of the gastrocnemius muscle lay close to 30° of plantarflexion with the knee extended, whereas the soleus muscle was maximally excitable close to 90° (or the position at which the sole of the heel is at right angles to the shaft of the tibia: the so-called "neutral angle"), indicating that for the soleus muscle, reflex excitability is reduced on either side of neutral, and for the gastrocnemius muscle, on either side of 30° of plantarflexion.

Burke, Andrews and Ashby (1971) studied the effects of static stretching on the "H" reflex (an electrically stimulated oligosynaptic reflex response) of the triceps muscle with the knee extended, and confirmed that stretches beyond neutral were reflex inhibitory. They also confirmed, by means of ischaemic studies below the knee and above the ankle joint, that the mediators of the inhibitory influence resided in the length-sensing muscle spindles within the muscles, rather than the tension sensing Golgi apparatus, cutaneous, or joint capsule receptors (see discussion).

This joint angle effect has been viewed as one of the confounding variables in assessing the monosynaptic tendon and H-reflexes in different pathological states, resulting in attempts at standardising methods of H-reflex assessment by excluding the joint angle effects to allow valid comparisons within and between subjects (Hugon 1973, see methods below).

Robinson, McComas and Belanger (1982) demonstrated that stretching the triceps beyond neutral produced a 46.9% (+/- 19.2% SD) reduction in H-reflex amplitude in all of the healthy subjects tested, whereas 5° to 15° degrees of plantarflexion beyond neutral augmented the H-reflex by 51.9% (+/- 42.1% SD) in 9 out of 10 subjects. Likewise, Tardieu and colleagues (1982) demonstrated an excitatory influence of the joint angle on the ankle plantarflexors when the triceps were stretched (dorsiflexed) from a position of relative plantarflexion through to neutral.

The apparent continuum of reflex excitability has since been confirmed in a number of experimental situations under which the muscles are active such as in treadmill walking and simple standing (Capaday and Stein, 1986) in which maximal soleus H-reflexes were obtained during the stance phase of gait, which is also the phase of gait during which the background electromyographic (EMG) activity is maximal. Similar results during the stance phase of treadmill walking were obtained by Crenna and Frigo (1987) who also looked at single limb treadmill walking and stepping on the spot to separate out the possible effects of a bipedal motor task. A further report by Capaday and Stein (1987) indicated that the central alpha-motorneurone drive such as during running could override the influence of muscle length in reflex modulation, because although the EMG output was 2.4 times greater in running than in walking, the H-reflex amplitude in running was either equivalent to, or often lower than that obtained in walking.

Gerilovsky, Tsvetinov and Trenkova (1989) studied the effects of electrode montages (monopolar and bipolar) as well as surface electrode positioning along the midline back of the calf, on the H-reflex amplitude. Although they were able to demonstrate variations in the absolute H-reflex amplitudes, depending on montage or electrode placement, the H/H-max ratio or relative change in H-reflex clearly altered with joint angle (muscle length) showing a 20-40%increase in the percentage H-reflex amplitude with the foot plantarflexed to -30° (120° absolute joint angle) compared to measurements at 0° (90° absolute joint angle) with the knee extended.

The greatest *relative* changes were captured by electrodes placed 1.5cm and 3.0 cm below the insertion of the gastrocnemius muscle whereas the greatest *absolute* changes were measured with electrodes placed 5.5cm and 7.0cm below the gastrocnemius insertion, however with each and every electrode placement, a larger H-reflex was obtained in -30° of plantarflexion compared to the neutral angle. Furthermore, identical relative recruitment curves were obtained irrespective of the use of a monopolar or bipolar electrode array with the calf muscle relaxed at an ankle joint angle of -10° (100° absolute joint angle) and the knee flexed to 30° (160°) from full extension. The authors state:

"For a given electrode location the amplitudes of the monopolar MU action potentials increased with an increase in joint angle (ie decreased soleus length) The degree of the amplitude increase of the MU potentials was comparable to that obtained for monopolar H-potentials."

Gerilovsky, Tsvetinov and Trenkova,1989.

More recently, Brooke and colleagues (1995) have indicated that H-reflex modulation which is maximum in stance and inhibited in swing, may be influenced by movement at the hip and knee. Simonsen and colleagues (1995) have reported a task-dependent aspect to H-reflex modulation, which is uniformly increased in stance in an uphill task compared with maximal at heel-strike in association with co-contracting muscles acting across the ankle in a downhill task.

Hultborn and colleagues (1996) have demonstrated that the inhibitory influence of static muscle stretches may last up to 10 seconds or more when the muscle is returned to its original length after an initial stretch. Further studies using trans-cortical magnetic stimulation (Hultborn *et al*, 1996), demonstrated that alpha-motorneurone excitability itself was *not* affected by muscle length, confirming the view that muscle length modulates presynaptic inputs to the motorneurone pool. In vitro studies by the same group demonstrated that the motorneurone input resistance and membrane potentials were not influenced by a dorsiflexion (static stretch) conditioning stimulus, indicating that the principal effect of muscle stretch beyond neutral is to produce a moderately long-lasting 'homosynaptic post-activation depression'.

7.2 Soleus muscle twitch characteristics, maximum voluntary contraction and joint angle.

Separate from the above studies on joint angle reflex modulation of recordable surface EMG potentials, the isometric mechanical torque of the muscles acting at the ankle joint have been extensively studied by Marsh *et al* (1981) for the tibialis anterior and Sale *et al* (1982) in the case of the plantarflexors in healthy adult subjects. Electrical stimulation studies have demonstrated that maximal torques are not obtained at the position of rest of the muscle *in vivo*, indeed, whether obtained as sustained maximal voluntary contractions (MVC), or indirect electrical stimulation of the muscle by its nerve supply, the tibialis anterior maximum torques were obtained near 10° of plantarflexion in the case of MVC or tetanisation at frequencies of 20-40Hz, whereas a 10 Hz frequency of electrical stimulation or single muscle twitches produced torque maxima at 30° of plantarflexion: ie in the almost fully elongated position of the muscle-tendon complex. Likewise, studies of the plantarflexors (Sale *et al* , 1982) indicated that soleus isometric torque maxima were obtained at 20° of dorsiflexion beyond neutral for muscle twitches, tetanisation at 10Hz and the MVC, with the knee flexed a 90°. The same group showed that plantarflexor torque output was greater with the knee

extended, when in addition to the soleus muscle, the gastrocnemius muscle is stretched (see Silverskjöld, 1923). Although the influence of knee extension was not as marked as the authors had expected, the maximum influence of the gastrocnemius occurring between 25° and 10° of plantarflexion: ie at a more plantarflexed joint-angle than for soleus muscle alone.

These combined studies suggest that the maximum torques at the ankle, developed by the dorsiflexors and plantarflexors, occurred when the muscle was stretched, and that the optimal angle did not correspond to that of the *resting in vivo length* as conventionally taught.

The present painless and non-invasive studies in adults and children look at the influence of static changes in the joint angle on mechanical reflex twitches obtained by tendon tapping at the ankle in terms of the reflex EMG amplitude, peak reflex twitch torques and twitch times with a view to correlating the electrophysiological and mechanical events.

The study aims to bridge the gap in our understanding of the joint angle on reflex electrical events, the ensuing reflex mechanical events and to contrast this with the effect of joint angle on the direct or axonally stimulated muscle. b).

7.3 Methods.

7.3 1 Mechanical equipment: design and arrangements.

In previous preliminary studies (see "Methods" in Walsh *et al*, 1993), subjects were seated with the ball of the foot on a 100kg load cell (RS Components). The system was isometric, but the initial load on the transducer varied with the exact placement of the foot on the transducer and the degree of relaxation Accordingly, the baseline from which measurements had to be made, varied continuously, so that to keep within the dynamic range of the instrument, repeated electrical rebalancing was required.

7.3.1.1 Adjustable high -inertia mechanical filter

An instrument capable of obviating the need for frequent electrical rebalancing of the force transducer was designed in the form of a high-inertia mechanical filter (figures 7.3.1a and b) comprising a 1m long beam coaxial with a large printed motor (G19M4, Printed Motors, Ltd, Bordon, Hants GU35 9HY) with a double shaft. A DC current up to 10A was passed through the motor which had a torque constant of $0.28NmA^{-1}$, ie the system being capable of exerting a maximum dorsiflexing torque of 2.8Nm. The dorsiflexing torque exerted by the printed motor being equivalent to the *baseline* or *resting tension* in soleus and Achilles

tendon, if no current was passed through the motor, or to the *incremental passive torque* at the ankle joint proportional to the tension in the soleus muscle and Achilles tendon, with increases in the current delivered to the torque motor. The beam was weighted at each end with 13kg lead weights to give the system high inertia designed to resist rapid plantarflexing or dorsiflexing torques: Brief torques generated by the ankle jerk affected the force transducer but were virtually over by the time the beam moved significantly. The lower end of the shaft of the motor was connected to a potentiometer to record the angular position in degrees (°)

7.3.1.2 High mechanical filter beam inertia.

The inertia of the system of 3.1kgm^2, was measured by applying a known torque and measuring the resultant acceleration.

7.3.1.3 Beam plantarflexor rotation following adult tendon taps.

Measurements of the plantarflexor rotation of the beam after a reflex twitch generated by tendon tapping in adults indicated that the beam rotated by only 0.035 rad or 2° by the time the relaxation was 90% complete. The deflection of the transducer when fully loaded with a 20kg force (196 N), which was more than twice the reflex force generated in any adult subject, corresponded to only 0.003 rad or 0.17° The arrangements of the apparatus thus ensured that measurements were virtually isometric.

7.3.2 Twitch force measurement.

The plantarflexor force was sensed by a 20kg load cell embedded in the beam and in contact with the ball of the foot (figs. 7.3.1 and 7.3.2), and the results converted to Newtons (N: maximum measurable force 196N): the raw data being expressed in Newtons and the normalised data for each limb in units of percentage torque (% Torque).

7.3.3 Electromyographic recordings and electrode placement.

The surface EMG was recorded from a bipolar arrangement of disposable silver-silver chloride electrodes (2.2x2.2 cm square Silver *Mactrode* , Marquette Electronics USA) with the active electrode placed in the mid-line posteriorly at the position of maximum calf diameter, the reference electrode placed distally just above the insertion of the muscle into the tendo Achilles, and the earth electrode on the knee-cap for each leg. This electrode positioning was chosen to simplify electrode placement between subjects of varying size, whose ages varied from 3 years to adulthood. For discussion of "ideal" electrode placement,

see Hugon (1973), "A discussion of the methodology of of the triceps surae T- and H-reflexes" in Desmedt (1973, pp773-780) and Gerilovsky *et al* (1989, discussed in section 4.1, above). The EMG signal was amplified, filtered at 20Hz-10kHz and stored on disc, along with the force and potentiometer signals on a *Medelec Sapphire 4ME* recorder (Medelec Limited, Old Woking, Surrey, UK).

7.3.4 Elicitation of soleus muscle reflex contractions.

Ankle reflexes were obtained in the resting muscle by manually delivering a blow to the Achilles tendon using a tendon hammer (fig. 7.3.1a and b and 7.3.2 , top) with an electrical switch in the head capable of triggering the recorder screen. In all cases four consecutive blows were delivered to the tendon with time between each blow to allow for reflex contraction and relaxation of the muscle. The results represent four tap-triggered averages for the reflex EMG and reflex muscle twitch at each given joint increment. Where possible measurements began at the *resting joint angle* which was always in plantarflexion (equinus) relative to the neutral angle, however in some individuals no reflex was obtainable until the soleus muscle had been stretched by applying a dorsiflexing torque with the motor.

7.3.5 Positioning of the subjects for study and determinations of the joint angle.

The experimental conditions for measuring a virtually isometric soleus muscle twitch following a tendon tap have been previously reported (Walsh 1992, Walsh *et al* 1993, Lin, Brown and Walsh 1994 and 1996b; Lin 1997 and Lin, Brown and Walsh, 1997).

Measurements were made with the subjects lying comfortably on the left side with the knee flexed to 90° (fig. 7.3.1a and b, fig. 7.3.2) to eliminate the effects of the gastrocnemius muscle which crosses both the knee and ankle joints (see Silverskjöld 1923, Sale *et al* ,1982). The ankle joint is coaxial with the axis of the horizontal beam which is free to rotate but heavily weighted at each end to increase the beam inertia, thus acting as a mechanical filter. The angular position of the ankle joint is monitored by a potentiometer zeroed to a 90° angle between the sole of the heel and the shaft of the tibia. The ankle joint position, and hence soleus muscle length was varied by means of a printed electrical motor coaxial with the beam capable of applying a graded torque up to a maximum of 2.8Nm (fig. 7.3.1a and b, fig. 7.3.2) so that each incremental joint angle was achieved at a known torque.

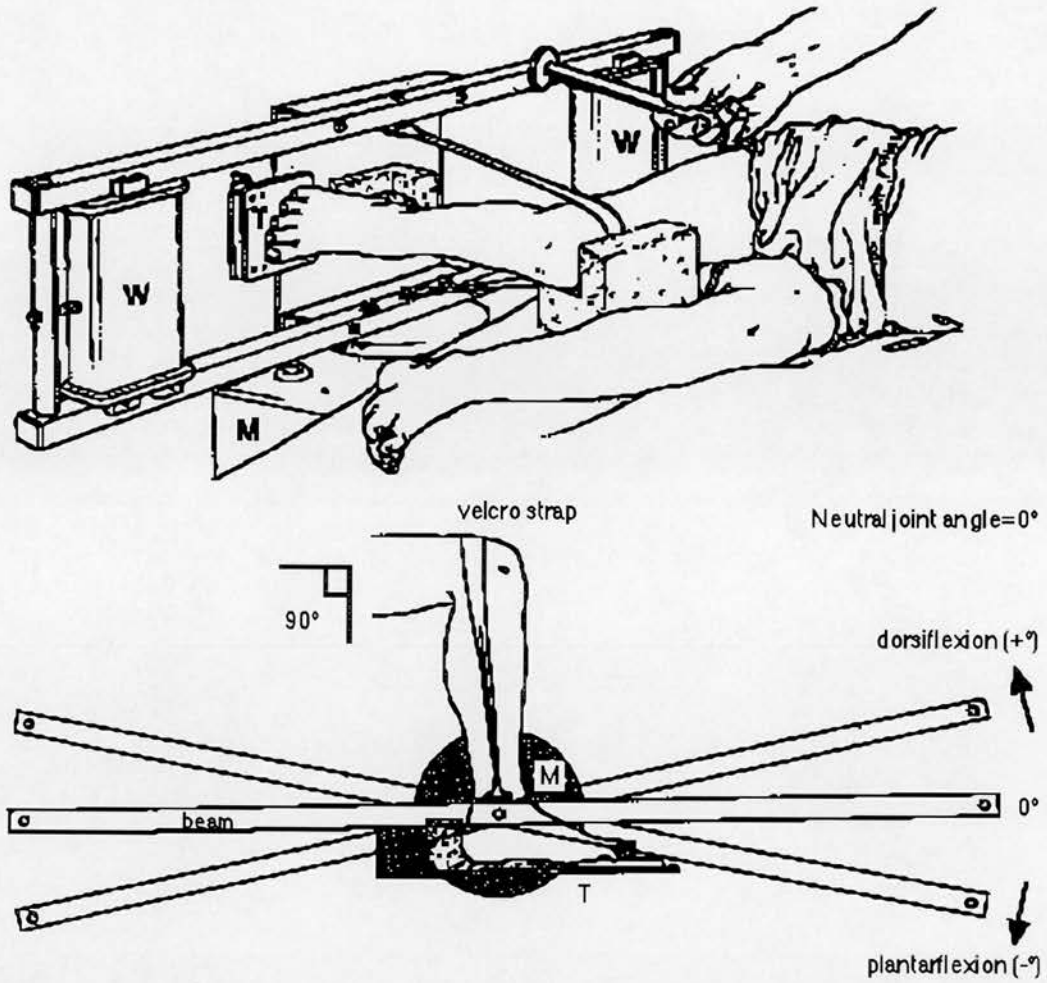


Figure 7.3.1(i). Diagram of high inertia beam, torque motor, force plate and limb positioning.
Arrangements for measuring near isometric soleus muscle reflex twitches.
Above: Artists impression of apparatus and **Below:** view from above. Note heavily weighted ends (W) of the beam, force plate (T), printed electrical motor (M) coaxial with the ankle joint. The subject lies in the left decubitus position to eliminate the influence of gravity. The hip and knee are flexed at right angles to eliminate the influence of the gastrocnemius muscle and the ankle joint is co-axial with the high inertia beam which is free to rotate. A printed electric motor, also coaxial with the beam and ankle joint applies incremental torques to dorsiflex the foot from the natural resting plantarflexion (negative joint angles) through neutral (0°, with the foot at Rt angles to the shaft of the tibia) into dorsiflexion (positive joint angles). The ball of the foot rests on the 20kg strain gauge plate to record the mechanical twitch generated by a tap to the Achilles tendon. The surface reflex EMG is recorded by disposable electrodes placed in the mid-line posteriorly at the position of maximum calf diameter for the active electrode and just above the point of insertion of the muscle into the tendon for the reference electrode. Measurements of four consecutive tap-averages were made at each increment of dorsiflexion maintained by the electric torque motor.
(**a.** reproduced with permission from Walsh 1992, *Muscles, Masses and Motion*, Mac Keith Press, **b.** after Lin, from Forssberg and Connolly, 1997. Drawings by Lesley Skeates-Bailey.

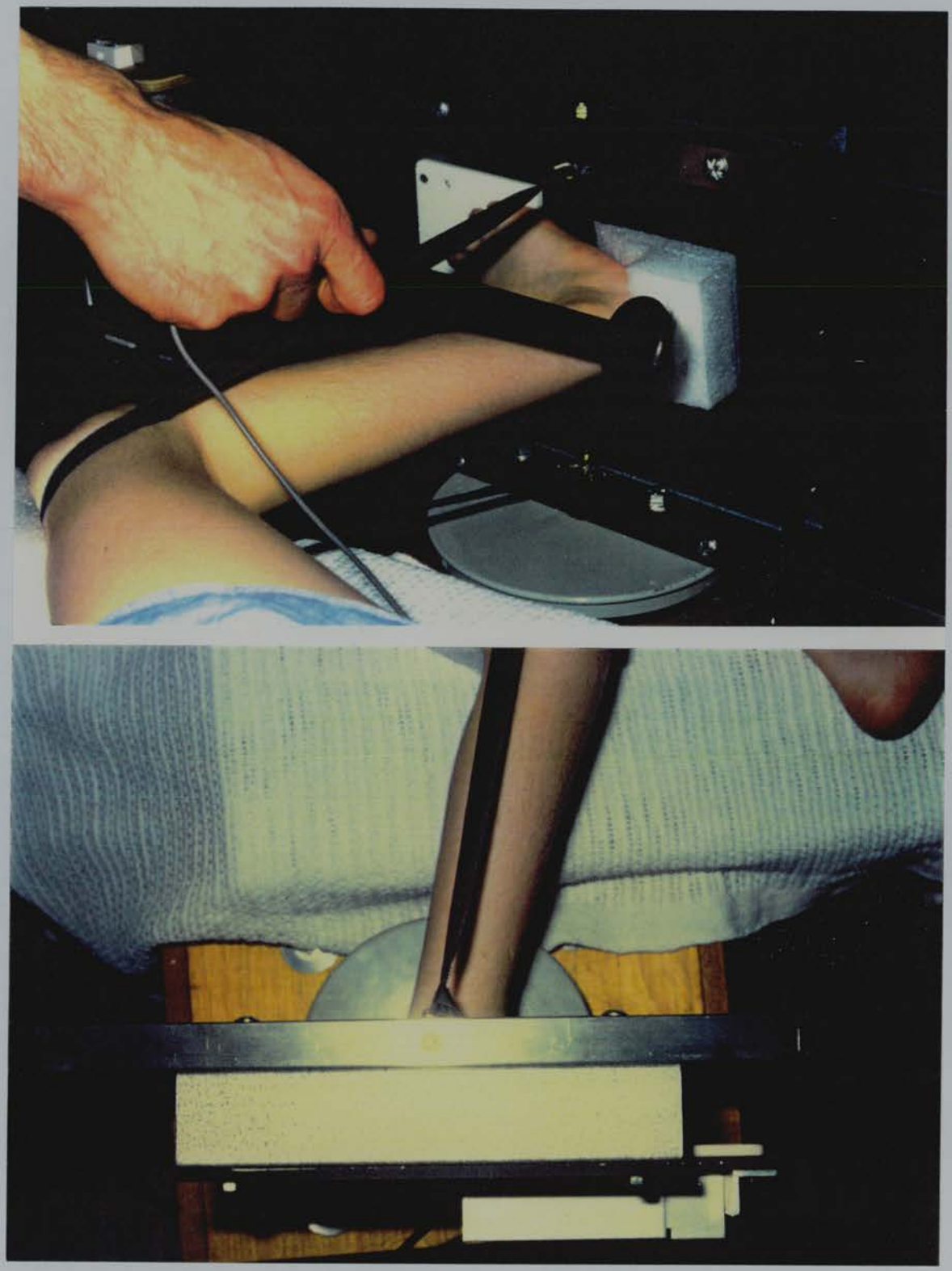


Figure 7.3.1(ii). Photographs of high inertia beam and experimental arrangements.
Above: Oblique view of high inertia mechanical filter, force transducer , co-axial torque motor, limb positioning and tendon hammer with electrical switch to allow tap-triggered averaging. (Surface EMG electrodes not shown and EMG recording equipment). Below: View showing the beam is co-axial with ankle joint. For details see figure 7.3.1(i) above, and text.

Throughout this study, *negative* angles refer to plantarflexion and *positive* angles to dorsiflexion beyond "neutral" respectively: "neutral" being the 0° angle, when the ankle is neither plantarflexed nor dorsiflexed with the foot at 90° to the shank. For each limb, reflex twitches were studied at small, sustained incremental torques of dorsiflexion from *resting plantarflexion* (with no passive dorsiflexing torque) through the neutral joint angle into maximum dorsiflexion.

7.3.6 Subjects.

The above apparatus and arrangements had previously been used to estimate the contractile properties of the soleus muscle in fifty-two male and forty-five female medical students (mean age 19.9+/- 2.4 years) and twenty-three male and seventy female older subjects (mean age male=68.3+/-4.8 years, mean female age 66.6+/-5 years): see Walsh, Wright, Davies, Lin and Thompson, 1993.

A convenience sample of thirty-one healthy controls were studied, nine adults aged 19-70 years and 22 children aged 3.9-13.6 years. All the studies took place in the same warm room under similar conditions. Parents gave informed consent and were present throughout for all children studied. A typical study lasted an hour for two limbs, with time to rest between limbs.Some limbs were studied twice to confirm reproducibility and reliability data.

7.3.7 Reflex twitches produced by tendon taps and electrical stimulation.

The characteristics of the reflex EMG and muscle twitch following a tendon tap is shown in figure 7.3.7a, illustrating the tap-triggered average of four consecutive tendon taps. The mechanical tap, seen as a brief stimulus artifact on the force trace (Newtons), causes a brief but large amplitude EMG discharge (mV) some 12-35 ms later, depending on subject age and limb length. This electrical discharge in the muscle is then followed by a slowly rising mechanical contraction, reaches a peak(PF) and then relaxes. The sequence of events: tendon tap, reflex EMG discharge and muscle twitch is referred to in the text as *neuro-mechanical coupling.*

In a few adult subjects, a fixed electrical current stimulus was applied percutaneously to the tibial nerve in the popliteal fossa to stimulate the soleus muscle via the motor axons, producing an EMG "M" response within a few milliseconds of stimulation at different joint angles (fig.7.3.7b), representing four consecutive electrical stimulus averages). By varying the electrical stimulus current (50-100mA, pulse width 100μs), the Ia afferents could reflexly

excite spinal alpha-motor neurones, producing the so-called Hoffman or "H" EMG response some 30-35ms later, depending on leg length and age of the subject. This latency is the time for the electrical impulse to travel from the point of stimulation up the 1a afferent (large diameter sensory) fibres to the spinal cord, depolarise the spinal alpha-motorneurones, travel back down the motor axons and depolarise the muscle (see figure 2.3 above for the anatomical route of the "reflex arc").

Direct stimulation by-passes the muscle spindle and the spinal cord apparatus: a supramaximal electrical stimulus would depolarise all the motor fibres contained in the nerve trunk while lesser stimuli would recruit varying numbers of fibres. This was done to compare the effects of the joint angle on reflexly stimulated soleus muscle output (via the spinal cord) with that produced by directly stimulating the axons supplying soleus muscle.

According to Hugon (1973), the gastrocnemius muscle H-reflex cannot be elicited when the gastrocnemius muscle is at rest, so that all the H-reflexes elicited in these studies are likely to be soleus muscle H-reflexes. Hugon (1973) has also stated that if the M and the H responses shared the same electrical waveforms, they were likely to originate from the same muscle: in the present case, the soleus muscle. In contrast to figures 7.3.7 a, figure 7.3.7.b shows three events on the EMG trace: **i**) an electrical stimulus artifact (arrowed, small downward deflection), **ii**) a large amplitude EMG discharge (M response) within a few milliseconds of the onset of the percutaneous electrical stimulus to motor axons of the tibial nerve, and **iii**) a small H-reflex about 30ms following the stimulus artifact. The force trace in figure 7.3.7 b shows no tendon tap artifact preceding the twitch, which instead is produced by the M response.

7.3.8 Quantitative analysis of electrical and mechanical events

The neurophysiological measurements were derived from traces obtained by four consecutive tap-triggered averages (see fig. 7.3.8.1), with time between taps for the electrical and mechanical events to elapse and for the instruments to reset: usually about 10 seconds.

Figure. 7.3.7. Soleus muscle tendon tap reflex and direct motor response. (overleaf).
a.(upper traces) The tap force artifact (N) is seen as a brief deflection on the force trace, followed 30-35ms later by the high amplitude reflex soleus EMG discharge (mV) on the EMG trace which lasts 5-8ms and is followed by the slowly rising and falling mechanical twitch force (N). The tracings represent 4 consecutive tap-triggered averages to compensate for variation in tap force between blows.

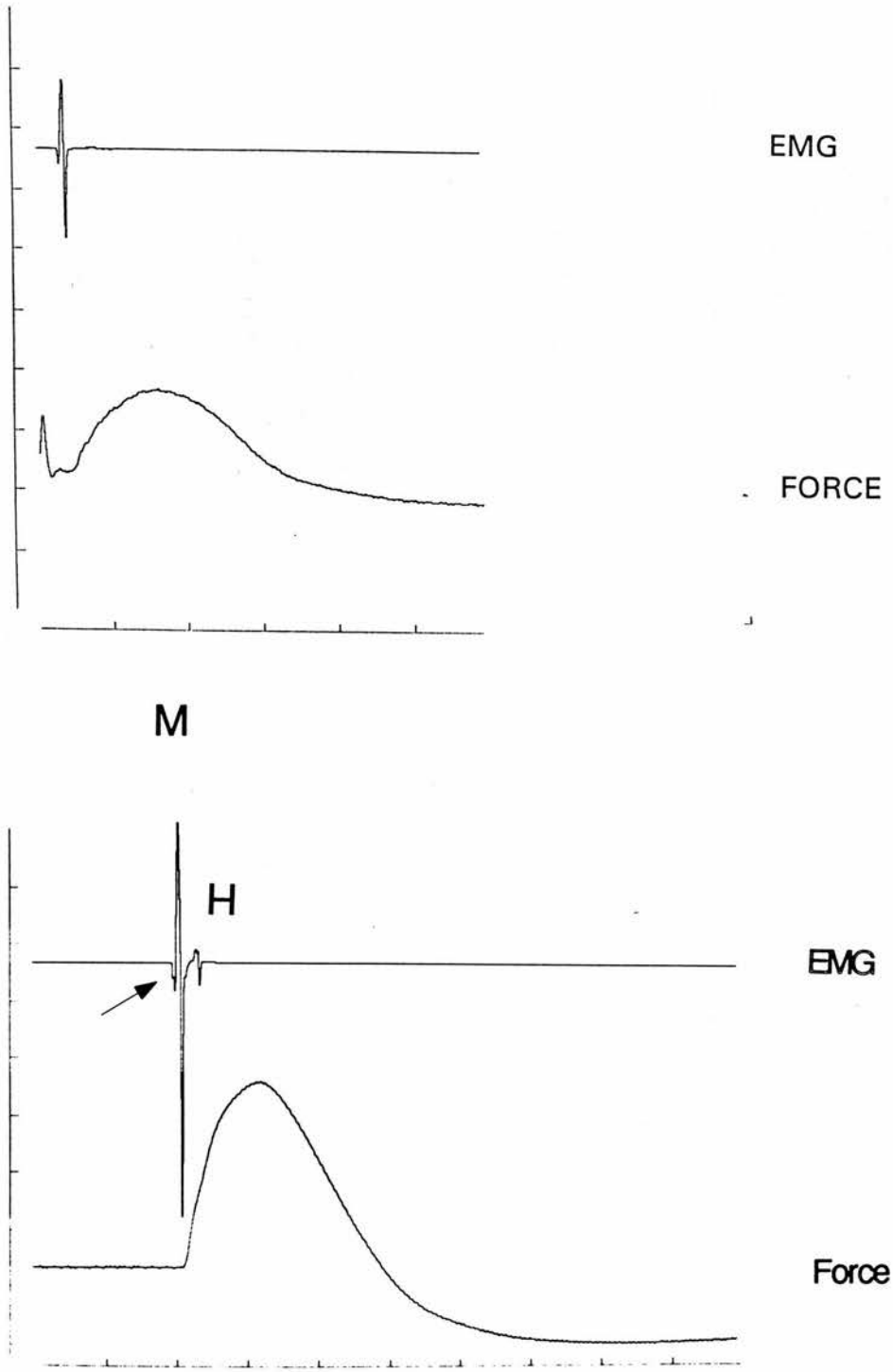


Figure. 7.3.7. Soleus muscle tendon tap reflex and direct motor response.
b.(lower traces) Percutaneous fixed current of 56mA with a pulse width of 100μs (brief downward deflection) on EMG trace and virtually coincides with the brief and large amplitude EMG discharge, the **M** response This is followed some 30-35 ms later by the onset of a brief and small amplitude Hoffman (H) reflex. Note similarity in M and H response waveforms. Irrespective of stimulus mode, mechanical twitch force (N) rises and falls slowly. Hor. int.= 100ms; vert. int.= 12.8 Newtons (force) and 2mV (EMG). 4 electrical stimulus averages.

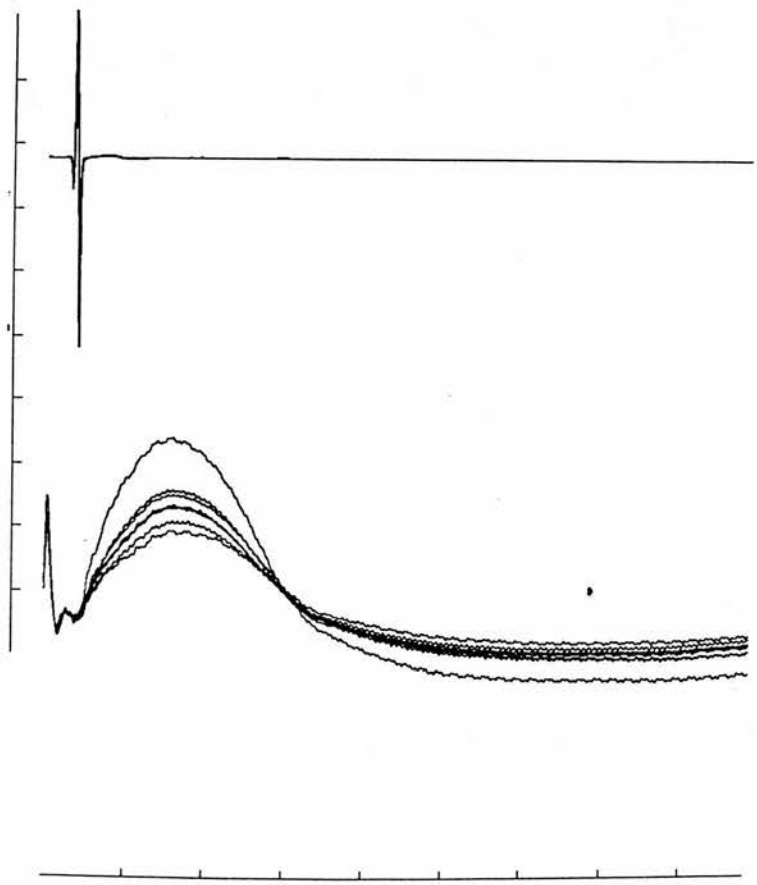


Figure 7.3.8.1. Superimposed single tendon reflex EMG and force traces
Eight consecutive tendon taps are superimposed. Adult male subject.
Top: Soleus EMG: triphasic waveform, vert. int.=2mV
Bottom: Isometric force traces, vert. int.=12.8N. Hor. Int.=100ms.

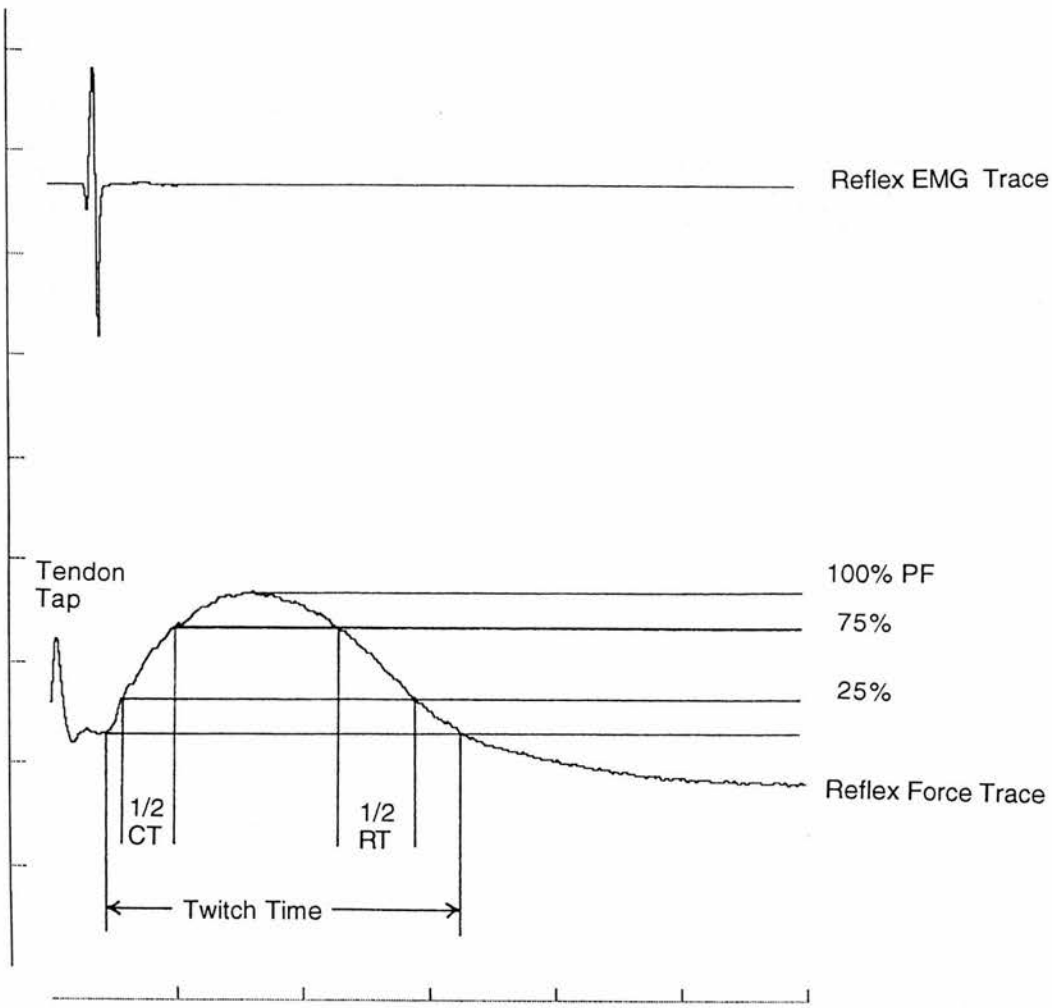


Figure 7.3.8.2 Quantitative parameters of soleus muscle reflex EMG and mechanical twitch.
Reflex soleus Tap, EMG and twitch measurements were derived from traces obtained by four consecutive tap-triggered averages with time between taps for the electrical and mechanical events to elapse and for the instruments to reset: usually about 10 seconds. Measurements included: tendon tap force in Newtons (N),or when normalised, the percentage tendon tap torque (% tap torque); the reflex soleus muscle peak to peak electromyogram (EMG) amplitude in millivolts (mV) or normalised soleus muscle EMG (%EMG); the absolute soleus reflex peak force in Newtons (N) or percentage units of torque (% Torque); the half contraction time (the inter quartile contraction time,1/2 CT, in ms); the 1/2 relaxation time (the inter quartile relaxation time, 1/2 RT, ms) and total twitch time (ms). Horizontal intervals= 100ms; vertical intervals= 12.8 Newtons for force trace and 2mV for EMG trace.

Figure 7.3.8.2 describes the the methods for measuring the reflex EMG and twitch characteristics. Measurements included: tendon tap force in Newtons (N),or when normalised, the percentage tendon tap torque (% tap torque); the reflex soleus muscle peak to peak electromyogram (EMG) amplitude in millivolts (mV) or normalised soleus muscle EMG (%EMG); the absolute soleus reflex peak force in Newtons (N) or percentage units of torque (% Torque); the half contraction time (the inter quartile contraction time,1/2 CT, in ms); the 1/2 relaxation time (the inter quartile relaxation time, 1/2 RT, ms) and total twitch time in milliseconds (ms).

7.4 Results.

7.4.1 Effects of the joint angle (soleus muscle stretch) on reflex twitch characteristics.

When the ankle reflex is elicited at sustained increments of dorsiflexion from the resting plantarflexor angle, through neutral, to maximum dorsiflexion, four phenomena are evident (figs.7.4.1a-b and 7.4.2 a-d, below).

7.4.1 a. Tendon tap studies and the joint angle.

i. EMG gain.

The reflex EMG and EMG gain (normalised reflex EMG/normalised tap torque ratio), reaches a peak and doubles close to -5° diminishing with further dorsiflexion beyond neutral till at +30°, the reflex gain is a fraction of the peak and resting values. It should be noted that no reflexes were obtainable in plantarflexion beyond the resting angle. The EMG and EMG gain diminish from +10° to +30° despite the fact that the tap torque (fig.7.4.2.1a) remains virtually constant at this joint range. The joint angle (muscle length) appears to modulate the recruitability of the motorneurone pool by initially *facilitating* and then *inhibiting* the reflex discharge.

ii. Torque gain.

The reflex mechanical gain (normalised twitch torque/ normalised tap torque ratio) steadily rises, increasing three-fold at +11° of dorsiflexion beyond neutral and then declining to original resting torques at +30° of dorsiflexion. It can be seen that the joint angle for maximum reflex mechanical gain differs from that of the maximum reflex EMG gain, the mechanical gain continuing to rise even though the EMG gain has started to wane. This indicates that there are mechanical factors which contribute to the reflex torque and which are capable of offsetting a drop in the number of reflexly recruited motor units (see fig.7.4.2.1) electrical stimulation below).

iii. Twitch time.

The twitch time remains constant at about 230ms from resting plantarflexion to +10° of dorsiflexion beyond neutral then steadily increases with further increments of dorsiflexion, reaching about 350ms at +30°, corresponding to an absolute increase of 130ms or 56% from that obtained in resting equinus (fig. 7.4.2.1).

iv. Twitch frequency.

The inverse twitch time or predicted twitch frequency declines from 4.25 Hz between -12° and +10° to 2.75Hz with dorsiflexion to +30°. This is the same as saying that soleus muscle tetanisation would be more easily achieved in full dorsiflexion than at the resting angle.

Legend to figure 7.4.1a-b Joint angle, the tendon reflex and direct soleus muscle activation.

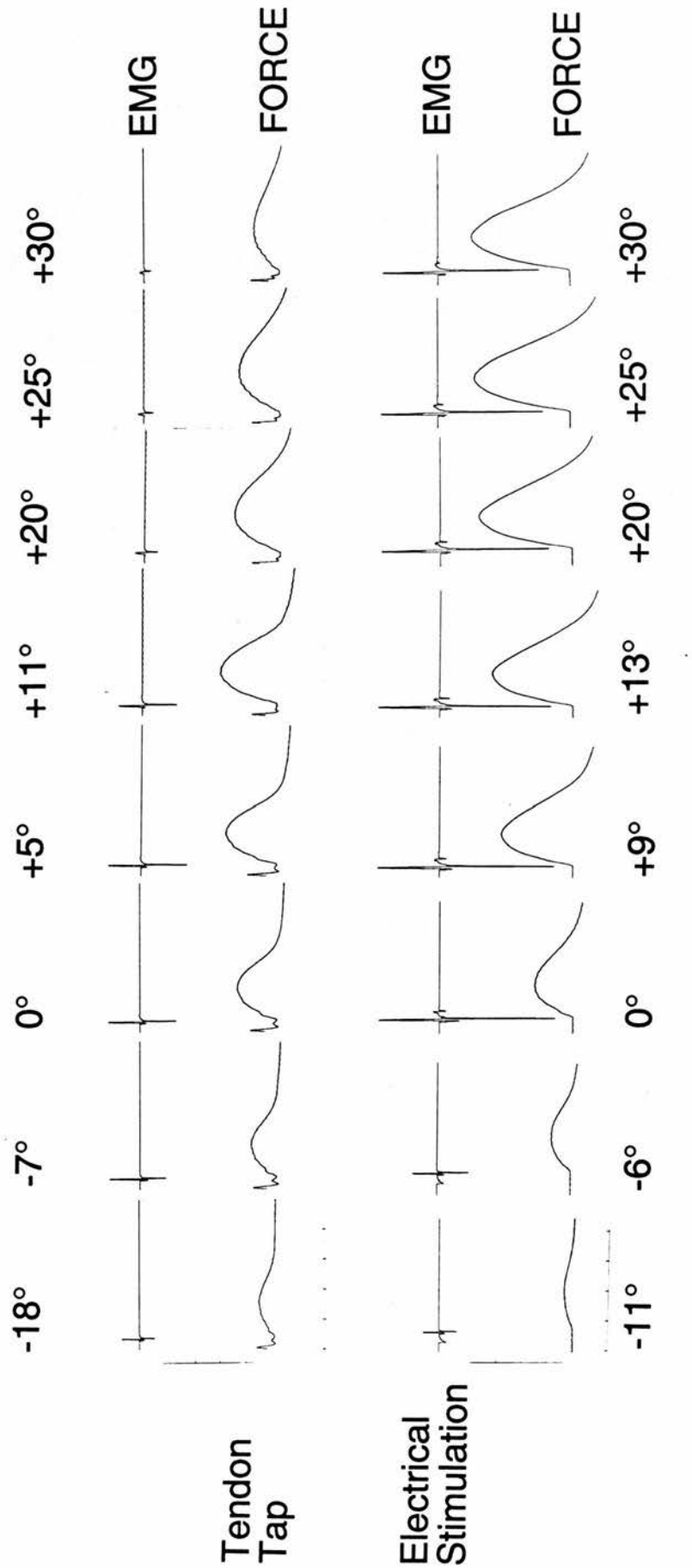
SEE LANDSCAPE FIGURE OVER LEAF

a. (above). Effect of joint angle on reflex EMG, twitch force and twitch time.
The reflex EMG (first series of traces) and peak force (second series of traces) become maximum close to neutral but wane rapidly in extremes of either plantarflexion or dorsiflexion. The muscle twitch time increases serially with each increment of dorsiflexion beyond neutral (see fig 7.4.2.1a-c).

b.(below). Percutaneous axonal electrical stimulation of the soleus muscle.
Same subject, experimental session, and electrode positions as the mechanical reflex studies in fig. a. above Each trace represents 4 electrical stimulus-triggered averages. At -11° and -6°, percutaneous axonal stimulation of the posterior tibial nerve produces a small electrical stimulus artifact (downward deflection) on the EMG trace followed by an H-reflex EMG discharge some 30-35 ms later. From 0° onwards the same electrical stimulus produces a large direct EMG discharge (the M response) which occurs within a few milliseconds of the electrical stimulus and is then followed by a small H-reflex some 30ms later reaching maximal amplitude at -6° (see figure 3 for clarification of M and H responses). EMG waveforms of the H and M responses are similar to the tap induced reflex EMG discharges of figure 5, indicating a common soleus origin (Hugon 1973). The M response varies little from 0° to +30° but the peak twitch force rises serially to a maximum at +30° (the optimal mechanical angle). By contrast the H-reflex diminishes beyond neutral.

Healthy adult age 31 years, Rt soleus, each trace represents 4 tap-triggered averages.
Horizontal intervals= 100ms;
Vertical intervals= 12.8 Newtons for force trace and 2 mV for EMG trace.

Figure 7.4.1a-b Joint angle, the tendon reflex and direct soleus muscle activation (legend: see above).



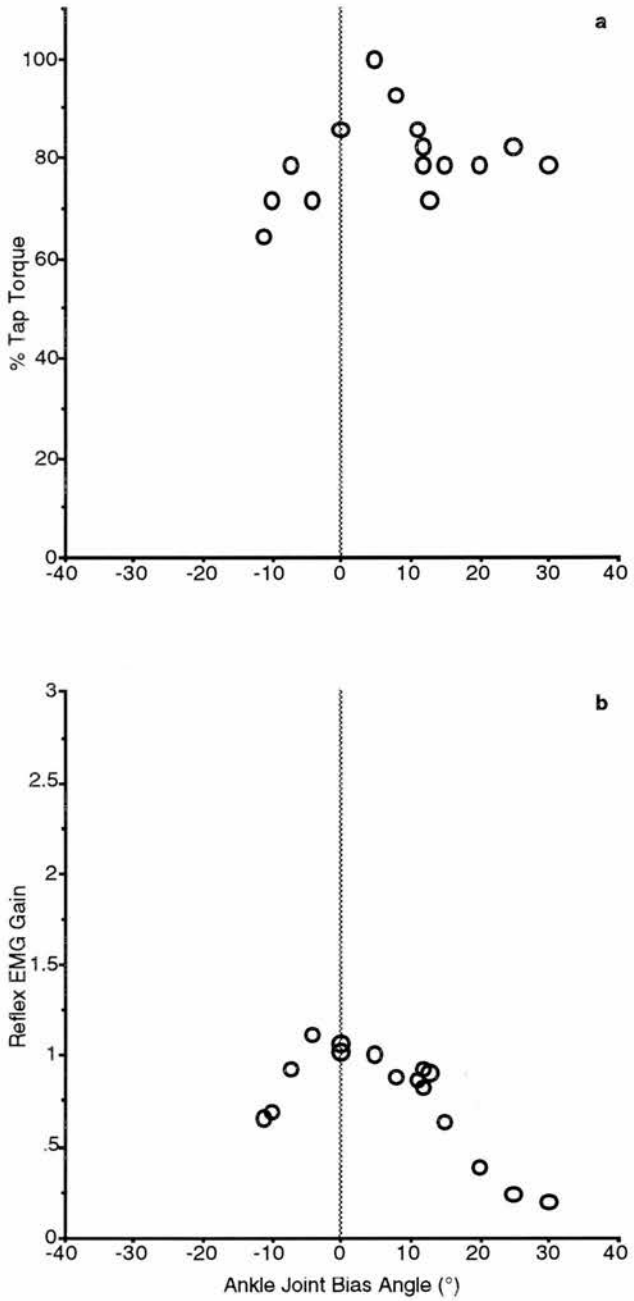


Figure 7.4.2.1a-b Reflex EMG and mechanical gain with changing joint angle.
Graphical representation of the data from fig. 7.4.1a.
a. Normalised tap torque (%Tap Torque) across the joint range. **b.** Change in reflex EMG gain (%reflex EMG/% tap torque) reaches a peak at -5° which is 63% greater than the resting gain and 450% greater than the gain at +30°
Rt leg, 31 year old adult male: each point =4 consecutive tap-triggered averages.0°= neutral angle: vertical dotted line in all graphs

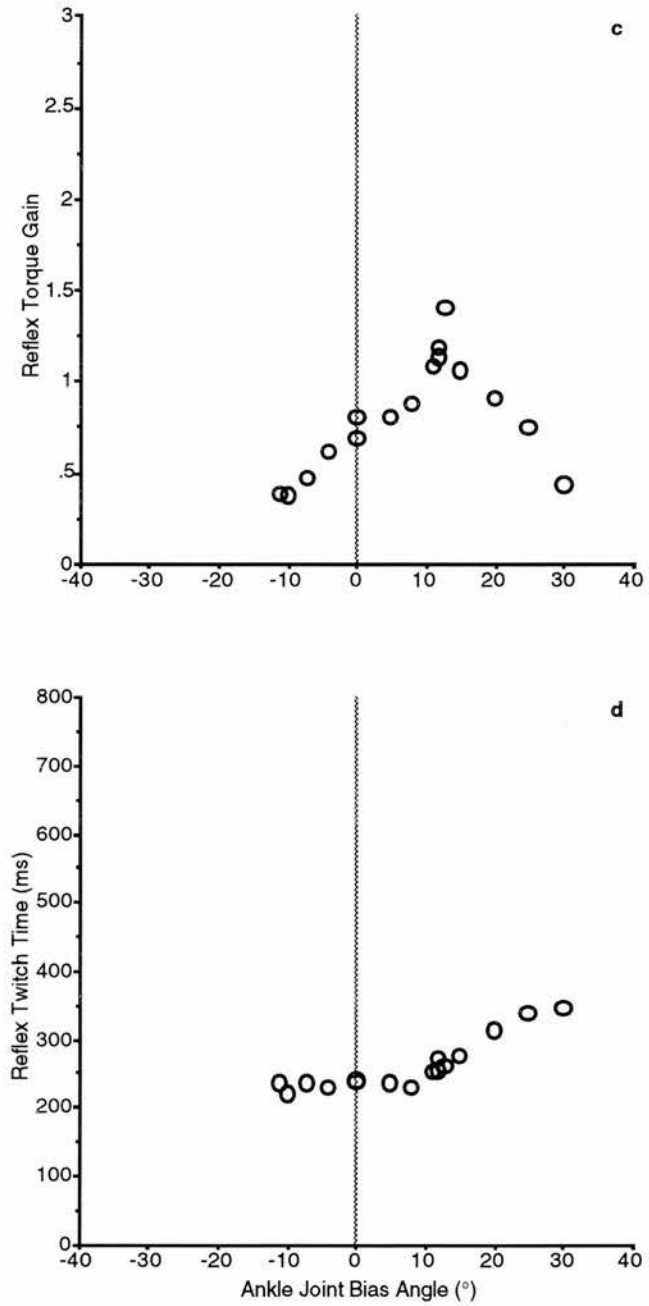


Figure 7.4.2.1c-d Reflex EMG and mechanical gain with changing joint angle.
Graphical representation of the data from fig. 7.4.1a.
c. Reflex mechanical gain (%peak twitch torque/%tap torque) against joint angle reaches a peak at +10° to +15° of dorsiflexion beyond neutral. There is a 265% difference between resting and peak mechanical gain and 221% fall in mechanical gain at +30°. The reflex mechanical output is maximal at the mid joint range, more than 20°dorsiflexed beyond the resting angle. **d.** Muscle twitch time (ms) remains constant at 225 ms from -12° (resting angle) to +10° increasing steeply to 375 ms at +30° of dorsiflexion beyond neutral.
Rt leg, 31 year old adult male: each point =4 consecutive tap-triggered averages.0°= neutral angle: vertical dotted line in all graphs

7.4 1b Electrical stimulation studies and the joint angle.

Figure 7.4.1b shows the results stimulus-triggered EMG discharge averages at different joint angles following four electrical stimulations of the soleus muscle via the posterior tibial nerve in the popliteal fossa (M- response) compared to the electrically stimulated monosynaptic H- responses and the accompanying muscle twitches (see fig.7.4.1a,b for explanation of M- and H- responses). At all joint angles the stimuli consisted of a fixed current of 56mA with a pulse width of 100μs. This electrically stimulated twitch data comes from the same subject as in figures 7.4.2.1 and 7.4.2.2 and was obtained at the same experimental session so that the conditions of electrical stimulation, including electrode placement, were similar to those for the tendon-tap reflex studies. Figure 7.4.2.2 illustrates these findings graphically.The soleus muscle is electrically stimulated at similar joint intervals to the tendon tap studies and the results can be summarised as follows:

i. Electrical events.

The electrical stimulus artifact (fig. 7.4.1b) is present on the EMG trace as the first brief downward deflection. At -11° and -6° of plantarflexion, stimulation of the posterior tibial nerve in the popliteal fossa produces an H-reflex EMG discharge some 30 ms later. From 0° onwards, the same electrical stimulus produces a large direct EMG discharge (the M response) which arises a few milliseconds after the electrical stimulus (figs.7.4.1b, upper traces and fig.7.4.2.2a). This M-response is seen as the largest deflection on the EMG traces from 0°, +9°, +13°, +20°, +25°, +30° of dorsiflexion, there is a small fall in amplitude of the M-response between +10° and +30°. Some 30 ms after the M-response is a small H-reflex EMG discharge (figs. 7.4.1b, upper traces and fig. 7.4.2.2b) which is maximal at -5° of plantarflexion and diminishes serially with dorsiflexion beyond neutral. Although the muscle spindles are by-passed by the electrical stimulus, the diminishing H-reflex with increasing dorsiflexion suggests that reflex alpha-motorneurone recruitment is inhibited by muscle stretch.

ii. Mechanical events.

When the muscle is stimulated directly along the motor axon and independently of the spinal alpha-motorneurone pool, the muscle twitch force (fig.7.4.1b, lower traces and fig. 7.4.2.2c) increases in strength to a maximum at +30°, indicating an apparent mechanical advantage of near-maximum dorsiflexion for the soleus muscle.

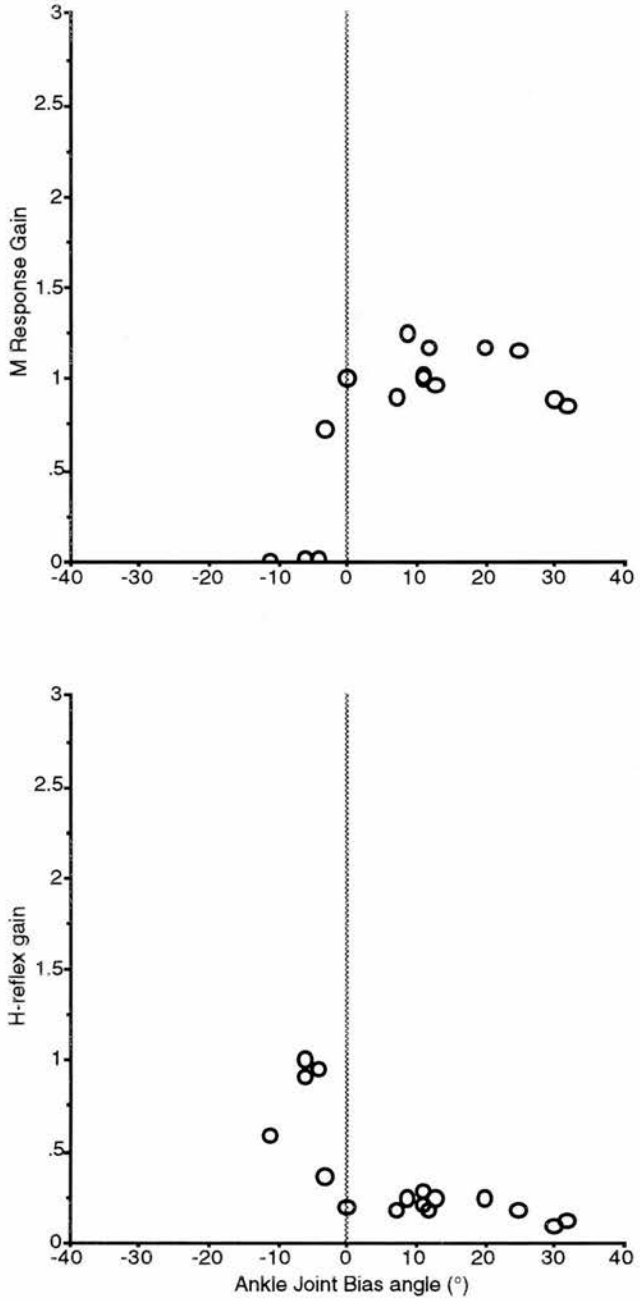


Figure 7.4.2 2a-b Percutaneous axonal stimulation studies.
Graphical representation from figure 7.4.1.b.
The M responses are compared with the Hoffman reflex (H-response) across the joint range. **a.** M-response gain (% M amplitude/% stimulus amplitude) remains reasonably constant after dorsiflexion beyond neutral **b.** H-reflex gain (%H amplitude/%Stimulus amplitude) wanes with dorsiflexion beyond neutral Rt leg, 31 year old adult male: each point =4 consecutive stimulus-triggered averages.0°= neutral angle: vertical dotted line in all graphs

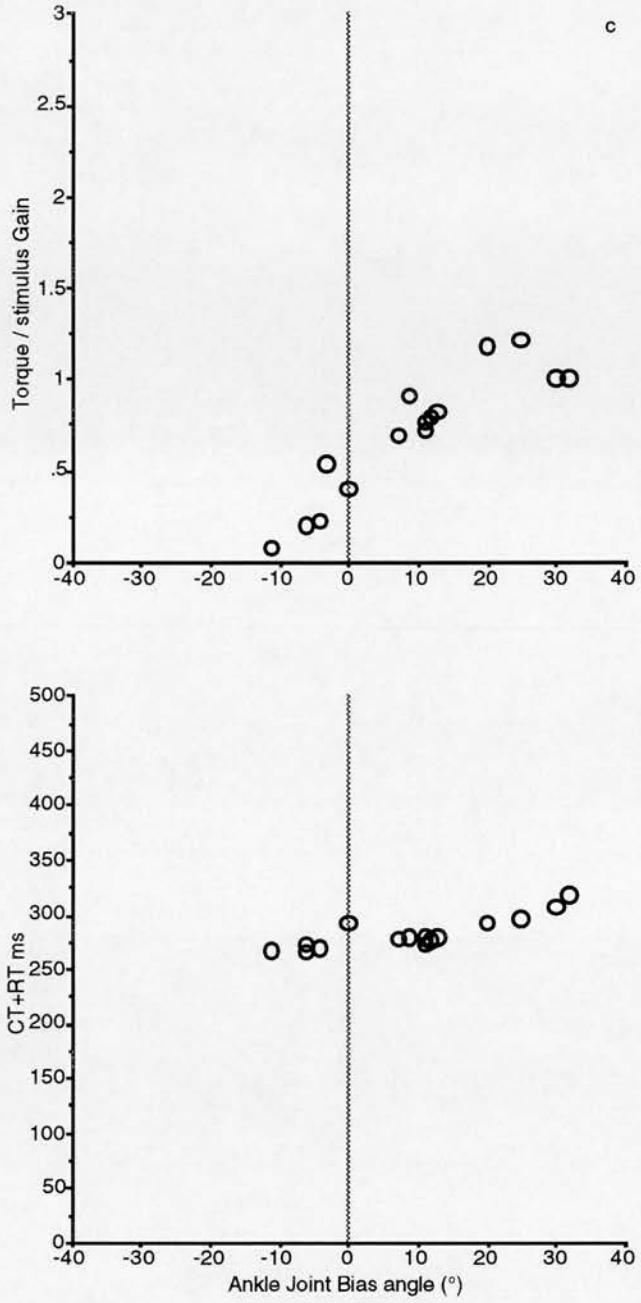


Figure 7.4.2 2c-d Percutaneous axonal stimulation studies.
Graphical representation from figure 7.4.1.b.
The M responses are compared with the Hoffman reflex (H-response) across the joint range.
c. Torque/electrical stimulus gain **d.** Twitch time (ms): this increases serially with increasing muscle stretch following the M twitches and appears to be independent of the mode of muscular activation, whether reflex or direct.
Rt leg, 31 year old adult male: each point =4 consecutive stimulus-triggered averages.0°= neutral angle: vertical dotted line in all graphs

If the motor axons are stimulated percutaneously, mechanical output of the soleus muscle increases with dorsiflexion through practically the whole joint range. This contrasts with the tap-induced reflex twitch force which reaches a peak at +11° beyond neutral and declines thereafter.

iii. Temporal characteristics.

The muscle twitch time increases serially with each increment of dorsiflexion (figs.7.4.1b, lower traces and fig.7.4.2.2d), apparently dependent on muscle length but independent of the mode of stimulation.

7.4.3. Varying electrical stimulation in equinus, at neutral and in calcaneous.

The responses of the soleus muscle to a fixed percutaneous electrical stimulus at varying angles have been studied above, suggesting an inhibitory influence of dorsiflexion on the H-reflex and a mechanical advantage to the soleus with incremental dorsiflexion.

The following study examines the response to a range of increasing percutaneous electrical currents (50-100mA, pulse-width 100μs) delivered to the tibial nerve in the popliteal fossa at the resting joint angle (-12° of equinus), at neutral and between +5° to +20° of dorsiflexion (calcaneus).

The stimulus intensity delivered was increased from 50mA in steps of 10mA to a maximum of 100mA. The data collected corresponds to four stimulus-triggered averages at each stimulus intensity. The subject was a healthy 25 year old male volunteer and recordings of the soleus muscle EMG and twitch were obtained as described in the sections above, in the relaxed state.

Figure 7.4.3.1 shows the polygraphic traces obtained at the various stimulation intensities and at the three joint angles described above.

Graphical data obtained from these traces are shown in figure 7.4.3.2a, the H-reflex to M- response EMG ratio was maximal at resting equinus, showing an obvious decline at neutral, declining further to 10-20% of the equinus value in calcaneus. These findings support those previously obtained with tendon-tapping studies which indicated an inhibitory influence of dorsiflexion beyond neutral on the reflex EMG amplitude.

Figure 7.4.3.2b shows the twitches obtained at different stimulus intensities and figure 7.4.3.2c indicates the soleus twitches attributable to H-reflexes and those to M-responses. In equinus, all the twitches were derived from the H-reflex. At neutral, the top two

stimulus intensities of 90mA and 100mA produced M-response twitches, while stimuli below these values resulted in H-reflex twitches. It is noteworthy that the H-reflex twitches at neutral are of greater magnitude than those obtained in equinus, despite the fact that the H-reflex is weaker at this joint angle than in equinus.

As indicated by the previous studies, the angle of maximal reflex excitability does not correspond to the angle of maximal twitch force: a weaker H-reflex producing a greater twitch in neutral as compared with equinus. In calcaneus, all the twitches are attributable to the the M-response alone: these M-response twitches being of greater force at equivalent stimulus intensities than any obtained at neutral.

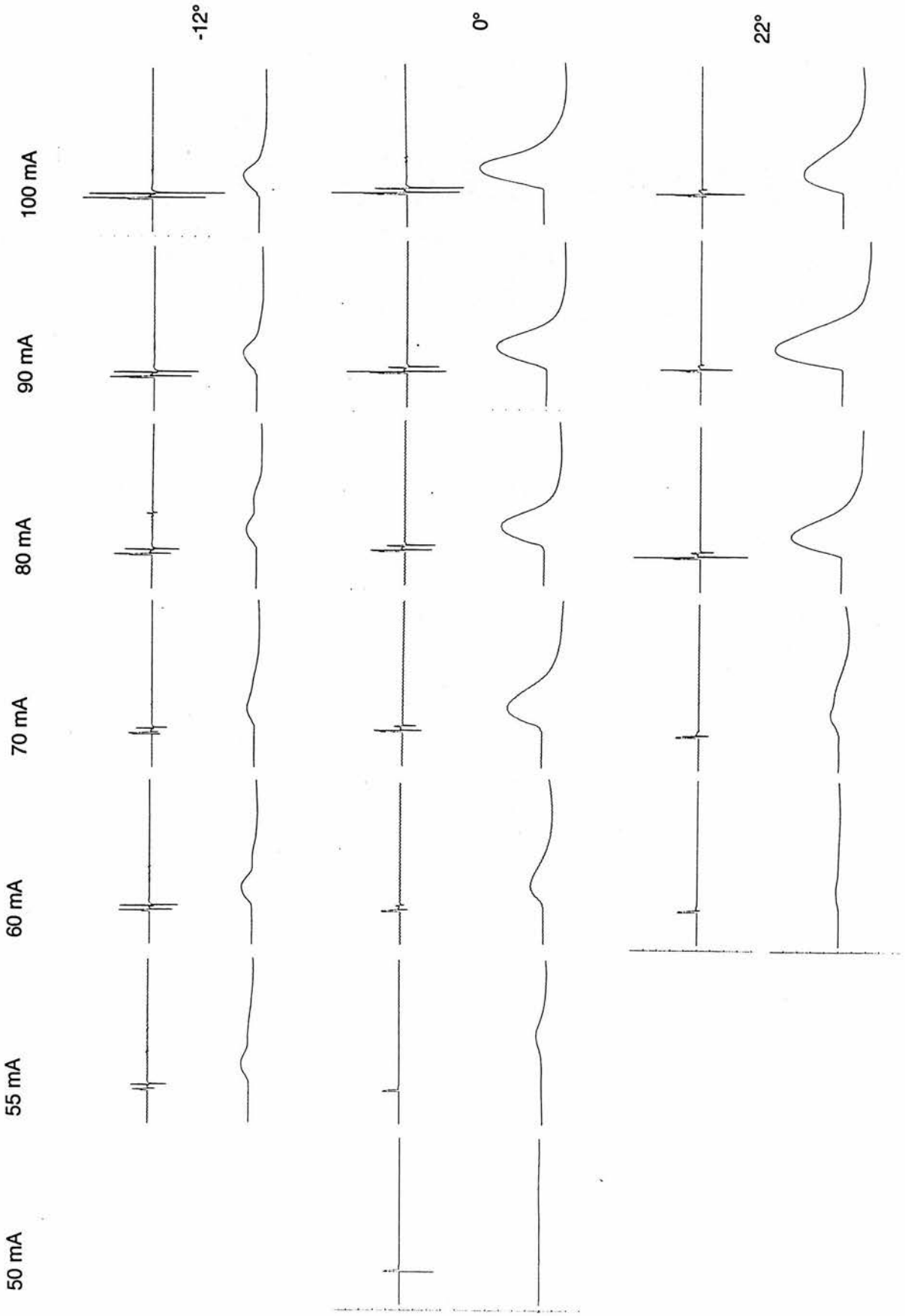
These studies seem to confirm the influence of the joint angle on reflex excitability, but there are a variety of possible explanations for the results:

i. The actual nerve stimulation intensities are different in equinus, at neutral and in calcaneus due to differences in proximity of the tibial nerve from the stimulating electrodes at the different joint angles. If this were the case, the current delivered to the nerve might increase with dorsiflexion, thus altering the H-reflex and M-response. However, the H/M ratio should not be affected by this (see discussion below).

ii. These results could be attributable to alterations in the motor neurone excitability with joint angle or indirectly to the level of presynaptic inhibition exerted on the motorneurone pool.

iii. The increasing mechanical twitches with dorsiflexion may in part be due to a combination of intramuscular and mechanical factors. Intramuscular factors include stretching of the in-series elastic slack component of the soleus muscle, alterations in the direction of pull of the muscle fibres and changes in the alignment of the actin-myosin cross-bridges at the differing joint angles. Mechanical factors include changes in the relative line of pull of the Achilles tendon, altering the effective bony lever arm with dorsiflexion.

Figure 7.4.3.1 Direct M and H-reflex twitch values at varying angles and electrical intensities.



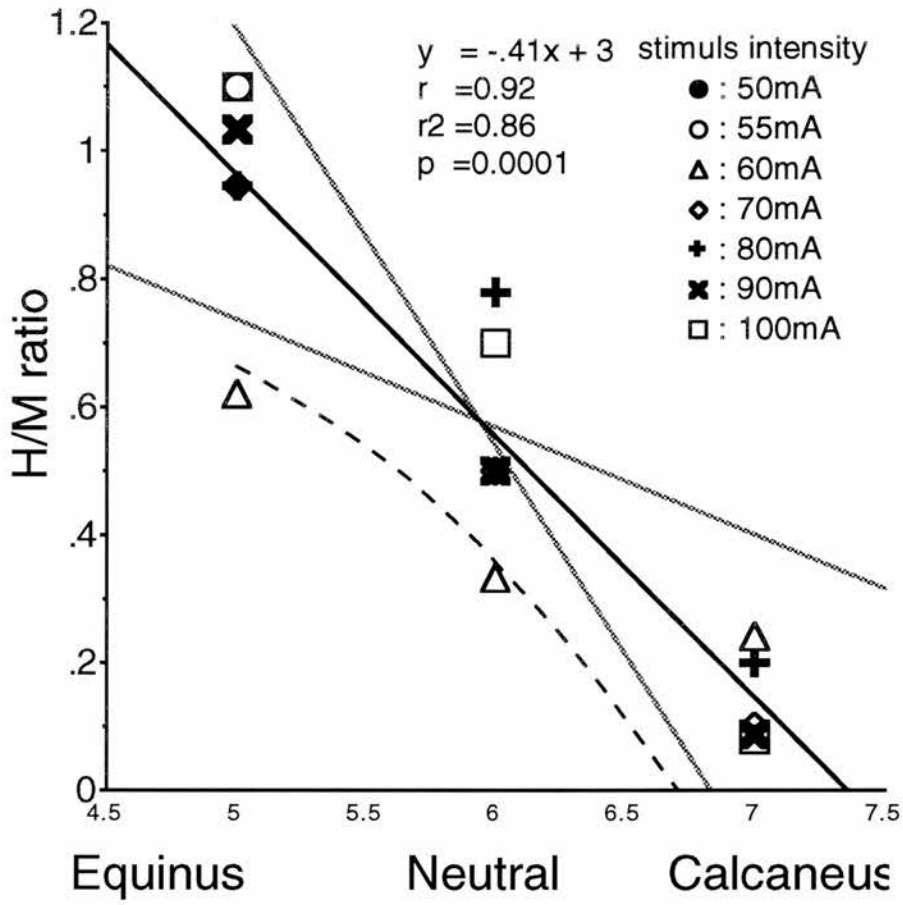


Figure 7.4.3.2 a. Soleus H/M ratio against joint angle.
Data obtained from figure 7.4.3.1
Electrical stimuli of varying current and fixed pulse-width of 100μs were applied percutaneously to the tibial nerve in the popliteal fossa of the right leg, resting healthy male subject, age 25 years. The subject lay in the right lateral decubitus position, with the hip and knee joints flexed to 90° (as for all previous and subsequent studies). The soleus muscle H-reflex and M-response EMG and mechanical twitches were recorded as described above in -12° of resting equinus, at neutral (sole of the foot at 90° with the shank of the leg) and in calcaneus (+5-22° of dorsiflexion beyond neutral). There is a clear decline in H-reflex to M-response EMG ratio with increased dorsiflexion, irrespective of the electrical stimulus strength. In calcaneus, the H/M ratio is 10-20% the value obtained in equinus. Note that the lowest stimulation intensities, 50-55mA, shown as open and closed circles, failed to produce either an H-or and M-response at neutral or in calcaneus, whereas in -12° of equinus, stimulation at 55mA produces a measurable H/M ratio.The mechanical H- and M-twitches are shown in fig.7.4.3.2 b and c, below.

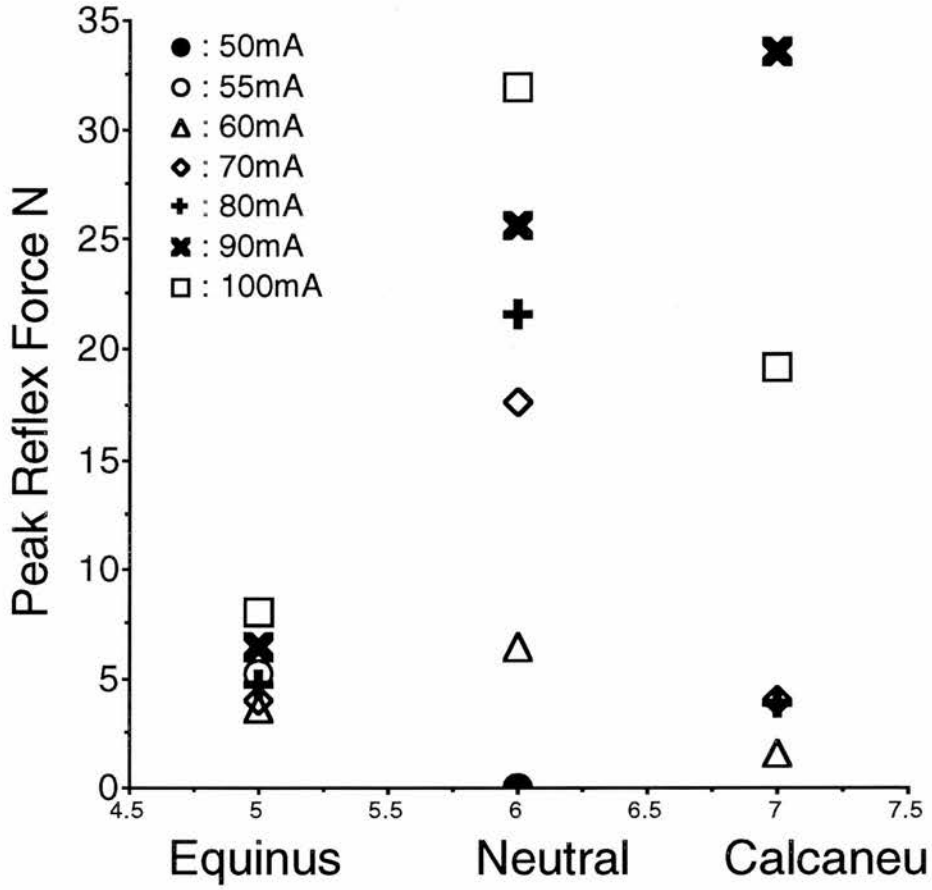


Figure 7.4.3.2 b Relationship between stimulus intensity and soleus muscle twitch force.
Data obtained from figure 7.4.3.1
The stimulus intensities at the three joint angles clearly produce differing twitch responses. Note that the twitches produced in equinus are tightly grouped, those at neutral and in calcaneus show wide variation in twitch strength.
Horizontal axis: Joint angle. Vertical axis: H and M twitch forces.
The distinction between the H-reflex and M-response twitches is shown in fig. 7.4.3.2 c, below. The experimental arrangements are as described in fig. 7.4.3.2a, above.

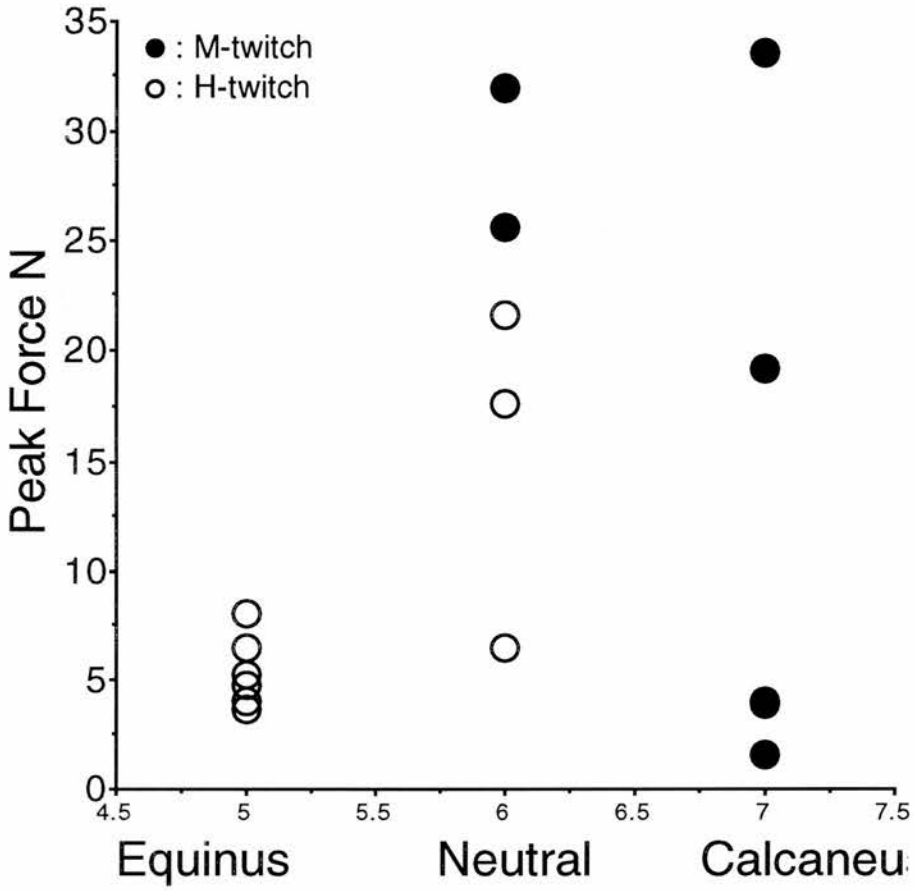


Figure 7.4.3c The effect of joint angle on soleus H-reflex and M-response twitches.
Data obtained from figure 7.4.3.1
The soleus twitches in equinus arise entirely from the H-reflex elicited at different stimulus intensities. At neutral, all but the pen-ultimate and maximum stimulus intensities produce H-reflex twitches and the strength of these has clearly increased with dorsiflexion. The experimental arrangements are as described in fig. 7.4.3.2a, b above.

7.4.4 Within and between subject comparative analysis.

The following studies explore whether or not the above findings can be generalised to subjects of different ages.

The means and standard deviations of the absolute reflex peak force (N) and reflex twitch time (ms) are plotted against age for the whole group. The tendon tap torques for each limb are normalised against the maximum value obtained to give the percentage tap torque which is plotted against the joint angle for each subject. The reflex EMG gain, defined as the normalised EMG to normalised tap torque ratio (% reflex EMG/% tap torque ratio), is plotted against the joint angle to examine the change in reflex gain against joint angle for all ages. Similarly, the reflex mechanical gain, defined as normalised peak reflex torque to normalised tap torque ratio (%reflex torque/%tap torque ratio), is plotted against joint angle. The absolute twitch times are plotted against joint angle.

7.4.4.1 Reflex EMG and mechanical gain with joint angle: adult group data.

Figures 7.4.4.1.2 a-c illustrate the effect of joint angle on the ankle jerk for nine healthy adults: nine right and eight left calves, each point representing four tendon tap averages. There is a clear variation of normalised reflex EMG and twitch torque with joint angle for the right and left soleus muscles respectively. In addition, the twitch time rises across the joint angle to a maximum in full dorsiflexion: for the group as a whole, the reflex twitch time varies from a mean of 265ms at -25° of plantarflexion, 267ms at neutral, and 383ms at +35° of dorsiflexion: a difference in twitch time of 118ms across the joint range or a 44.5% increase with dorsiflexion.

Figure 7.4.4.2a shows variations in normalised tendon tap torque across the joint range for the 17 adult soleus muscles. The group data confirm that the maximum *reflex EMG gain*, defined as (reflex EMG/tap torque ratio) occurs from -15° to -10° plantarflexion (fig.7.4.4.1.2b) in contrast to the *reflex mechanical gain* (reflex torque/tap torque ratio) which is maximal between 0°and +10° of dorsiflexion (fig.7.4.4.1.2c) indicating a net gain in reflex torque for any given tendon tap modulated by the joint angle. The angle for maximum reflex EMG gain differs from that of the resting joint angle, and the maximum reflex mechanical gain is obtained with a further dorsiflexion of 20° to 25°.

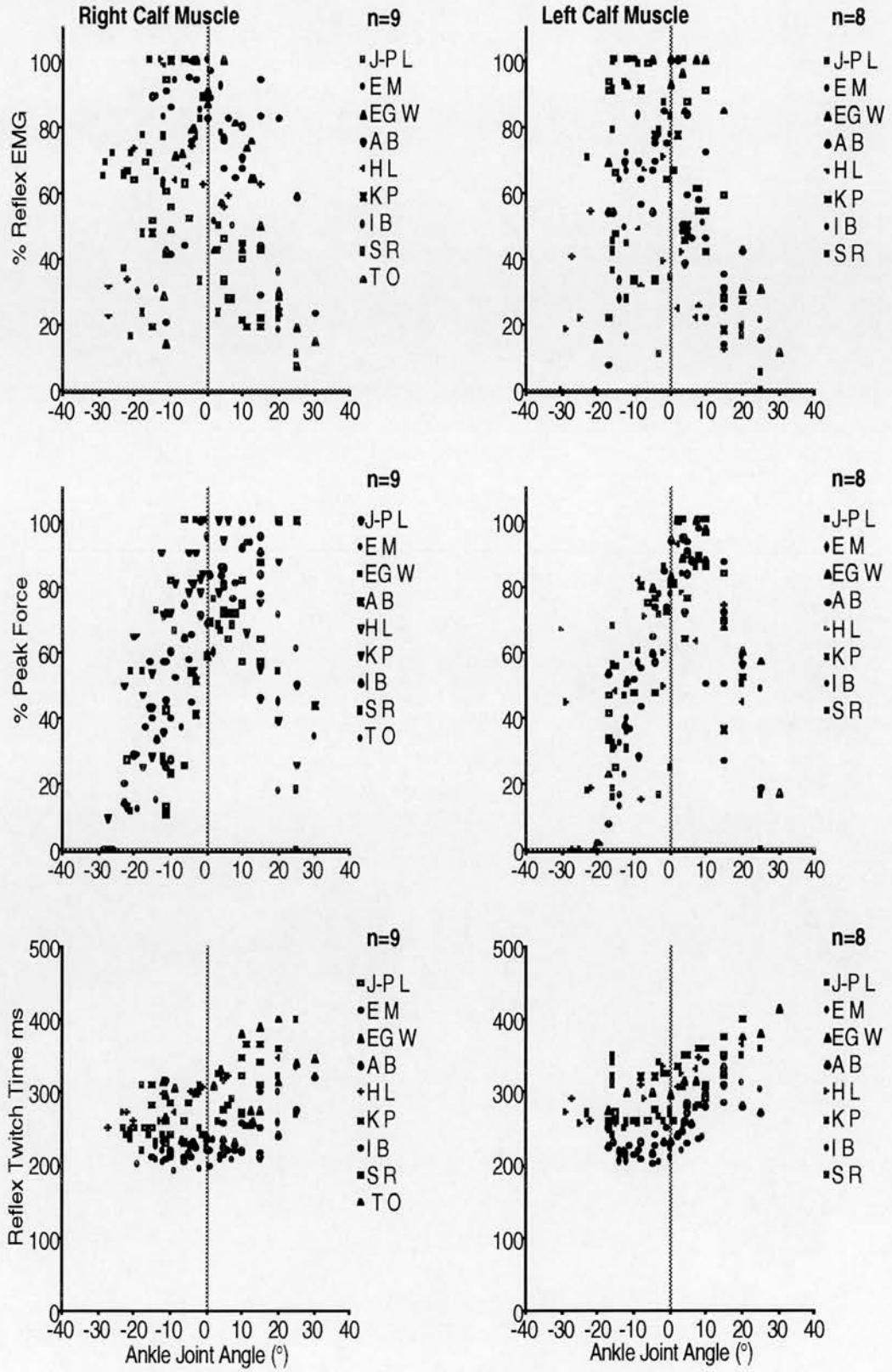


Figure 7.4.4.1 Combined tendon tap reflex data nine healthy adults
The data were obtained for 9 right and 8 left soleus muscles across the joint range. Hor.. axis=ankle joint angle(°), Vert. axes= reflex %EMG,%Force and Twitch time.

Legend to figure 7.4.4.1.2 a-c. The effect of the joint angle on adult reflex excitability.

see over leaf.

a. Normalised tendon tap stimulus (% Tap Torque) across the joint range
b. Reflex EMG gain (% reflex EMG/%tap torque) is maximal between -15° and -10° of plantarflexion
c. Reflex torque gain (%reflex torque/%tap torque) is maximal between 0° and +10° of dorsiflexion,
9 healthy adults age 19-70 years: 9 Rt and 8 Lt legs, 6 males and 3 females.
Each point represents 4 tap-triggered averages.

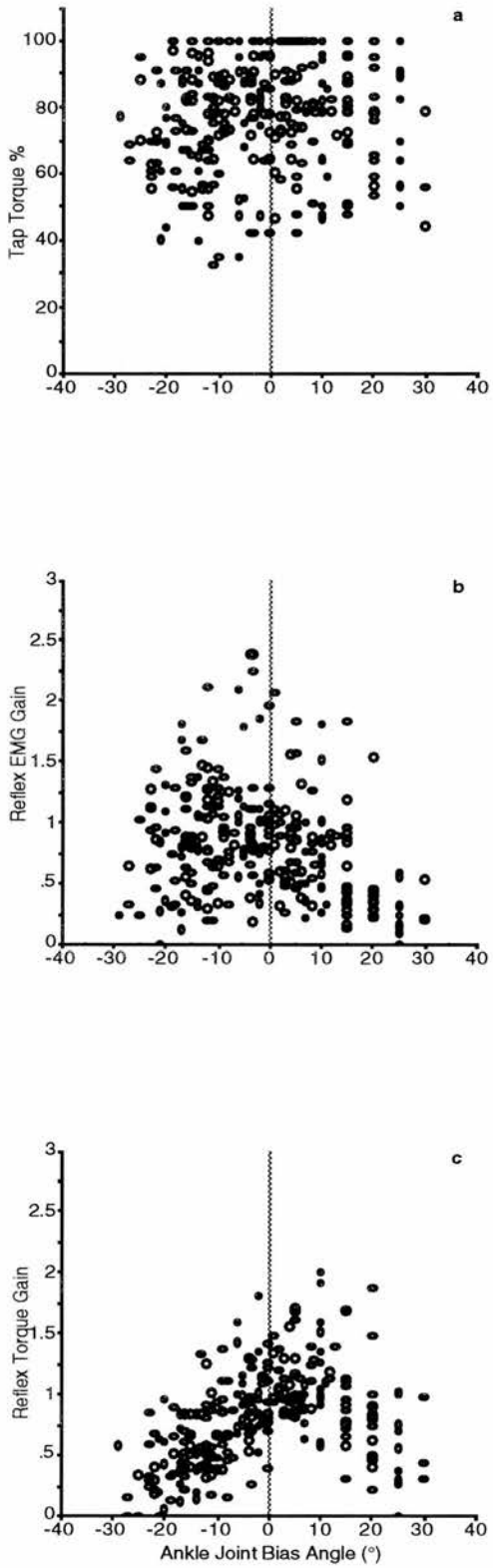


Figure 7.4.4.1.2 a-c. The effect of the joint angle on adult reflex excitability.

7.4.5 Effects of age on soleus muscle biomechanical and reflex twitches in children.

The effect of age on the biomechanical and contractile properties of muscle is demonstrated in figure 7.4.5.1 a-c, which depicts the mean and standard deviation across the joint range for the 31 subjects who underwent tendon tapping studies: **a**. soleus muscle compliance (°/Nm), **b**. the applied dorsiflexing torque (Nm), **c**. the reflex peak force (N) and **d**. the reflex twitch time (ms). The mean peak reflex force (N) increases five to eight-fold with age, whereas the mean twitch time diminishes by 30% from 325 ms to 225 ms between 3 and 19 years, slowing again thereafter for the seven subjects over 19 years. The corresponding mean twitch frequency (inverse twitch time, not shown) increases from 3Hz to 4.5 Hz.

Figure 7.4.5.2 a-d shows the reflex data for the soleus muscles of 22 children aged 3.9 to 13.6 years old. The data represents subsets from 41 soleus muscles, 22 right and 19 left respectively, one left limb being untested in a three year old girl to shorten the test time. In two cases, no reflexes were obtainable on the left side after having tested the right.

The paediatric data is more scattered than that of the adults. Nevertheless, the maximum reflex EMG gain is skewed to between -15°and -10° of plantarflexion (fig. 7.4.5.2b) whereas the reflex torque gain is maximal between 0° and +10° of dorsiflexion (fig. 7.4.5.2c). The reflex twitch time, is likewise more scattered, but increases with dorsiflexion, rising from a mean of 277ms at -25° and 285ms at neutral to 370ms at +35°: a mean difference of 93ms across the joint range, or a 33.5% increase. As figure 7.4.5.1d illustrates, the slowest twitches occur in the youngest children, reaching young adult values by the age of 10 years. The paediatric and adult reflex data are similarly influenced by the joint angle.

These experiments indicate that short-term muscle stretch modulates reflex excitability, presumably by altering presynaptic input to spinal motorneurone.

This demonstrates the effect of limb posture on the expression of the excitability of the nervous system.

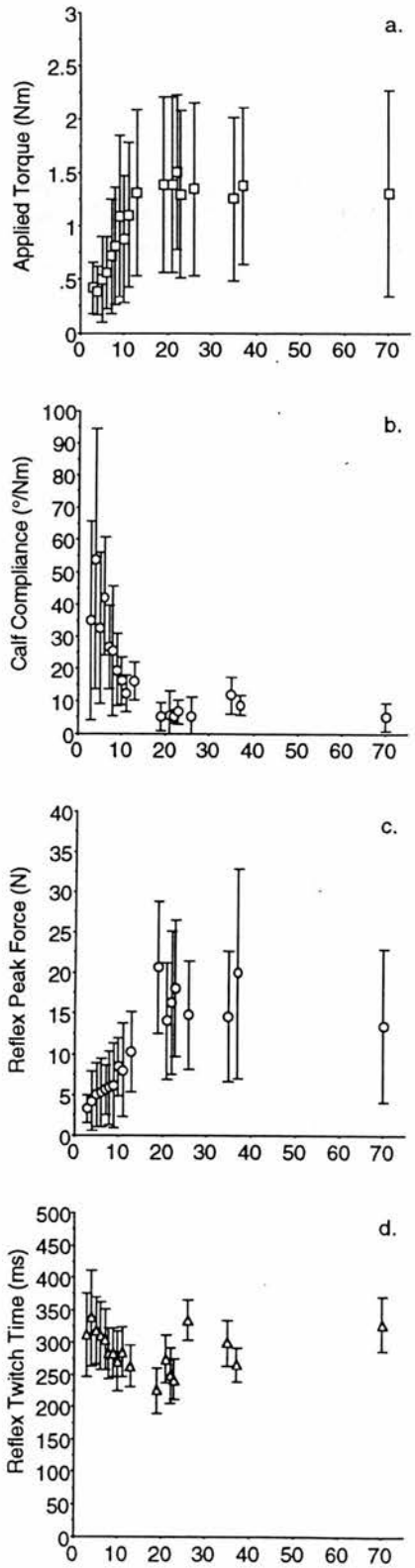


Figure 7.4.5.1 a-c Changes in soleus muscle characteristics with age.
Mean +/-SD for 31 healthy subjects aged 3.9-70 years.
a. Applied dorsiflexing torque (Nm) rises.
b. Soleus muscle compliance (°/Nm) diminishes in the first decade of life.
c. Reflex peak twitch force (N) increases.
d. Reflex twitch time (ms) falls over the same period.
from Lin, Brown and Walsh, 1997. By permission.

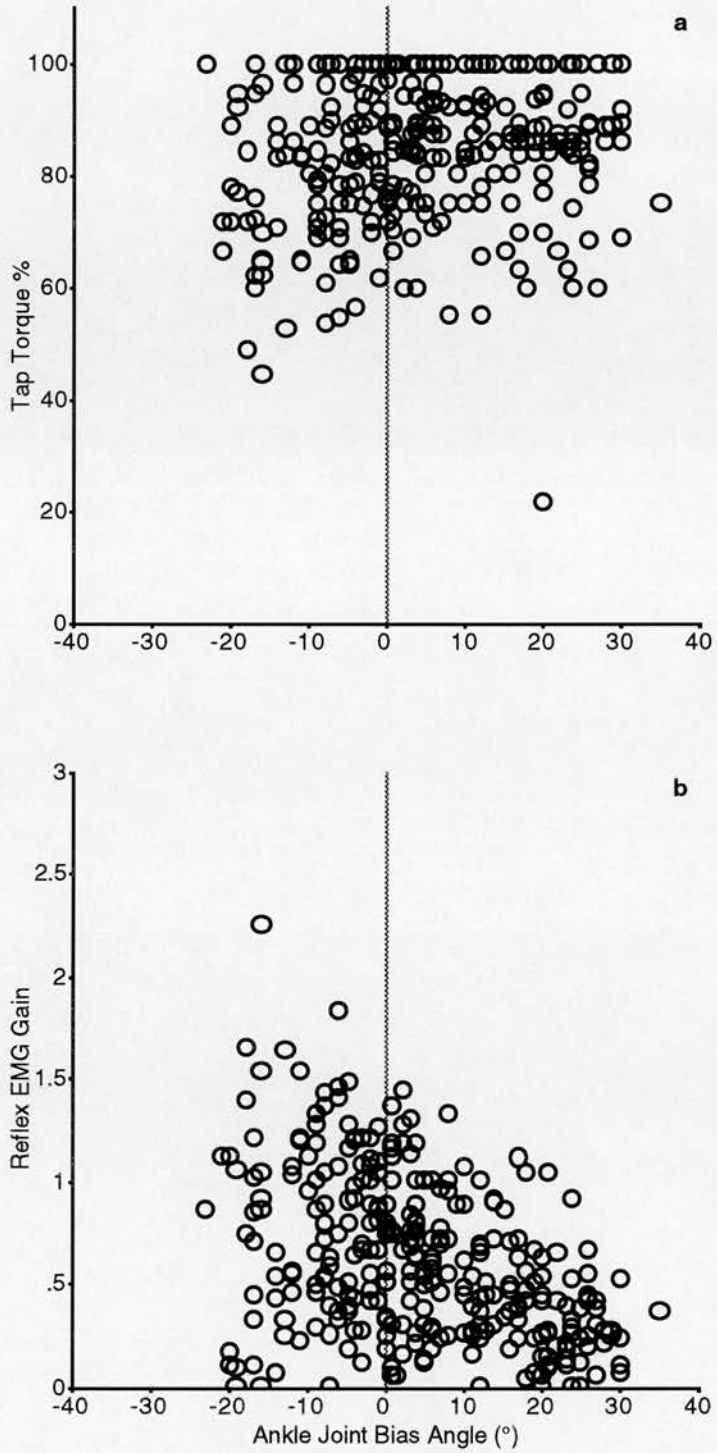


Figure 7.4.5.2 a-b Paediatric reflex twitch data against joint angle
a. Normalised tap torque (% tap torque): stable throughout the joint range
b. Reflex EMG gain (reflex EMG/Tap torque ratio): rises to maximum in plantarflexion
Reflex data for the soleus muscles of 22 children aged 3.9 to 13.6 years old. The data represents subsets from 41 soleus muscles, 22 right and 19 left respectively.Each point is the sum of 4 consecutive tap-triggered averages.

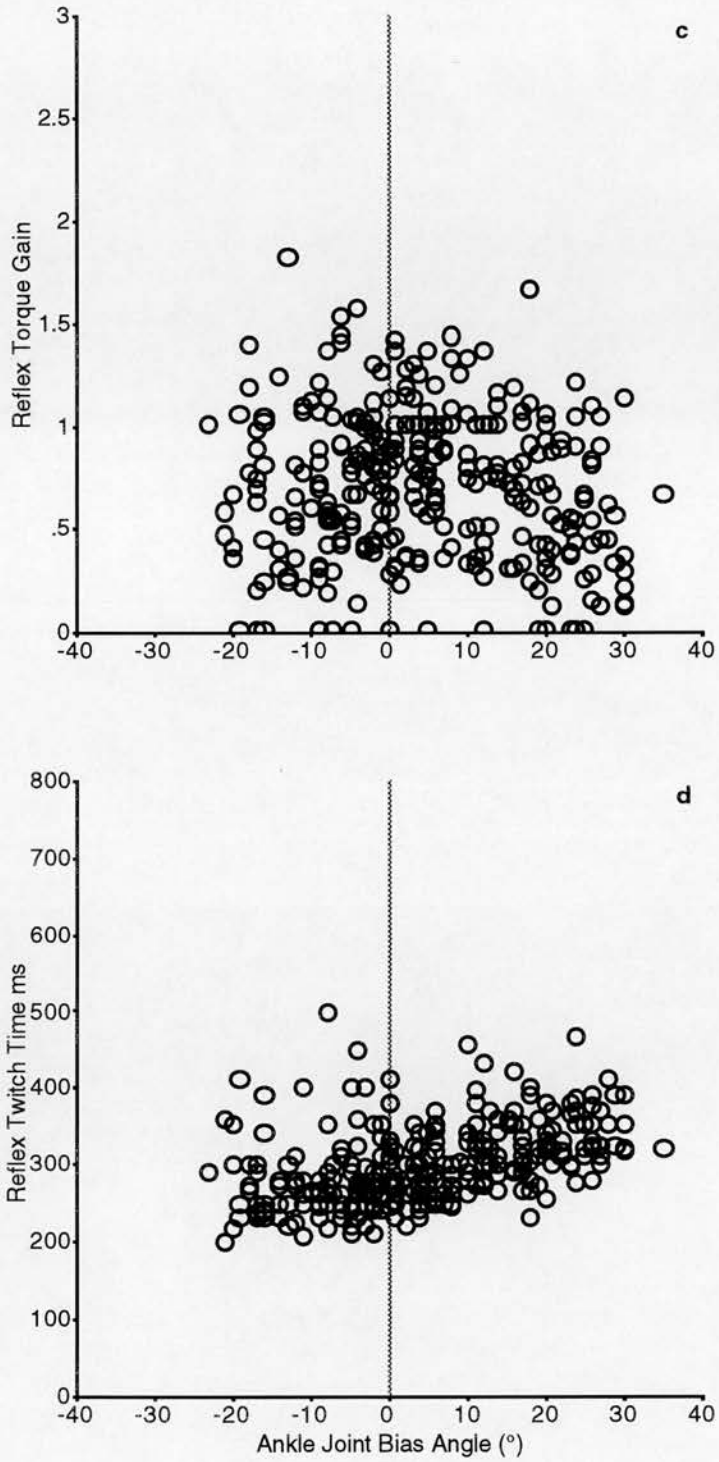


Figure 7.4.5.2 c-d Paediatric reflex twitch data against joint angle

c. Reflex twitch gain (reflex twitch torque/tap torque ratio): the maximum is in dorsiflexion

d. Reflex twitch time(ms): increases serially with dorsiflexion ie muscle lengthening.

Reflex data for the soleus muscles of 22 children aged 3.9 to 13.6 years old. The data represents subsets from 41 soleus muscles, 22 right and 19 left respectively

7.4.6 Summary of results for adults and children.

The influence of the joint angle on stretch-reflex excitability of the soleus muscle at the ankle has been studied in 22 children aged 3.9-13.6 years and 9 adults aged 19-70 years.

7.4.6.1 Joint angle and reflex EMG gain.

For all subjects, reflex EMG and mechanical twitch torque gain were trivial at *resting plantarflexion.* The *reflex EMG gain* reached a maximum between -15° and -10° of plantarflexion beyond the neutral angle, 0°, defined as the foot at right angle to the tibia, diminishing steeply with further dorsiflexion. These studies confirm that the in *vivo* excitability of the spinal alpha-motorneurone pool is strongly influenced by muscle length and explain the variability in reflex excitability within and between subjects, if the joint angle is not controlled. They also indicate how posture may influence force production, and hence movement, agreeing with the known function of the soleus muscle in the stance phase of gait and the modulation of motor unit recruitment during voluntary alternating movements at the ankle (see section 9 below on alternating movements).

7.4.6.2 Joint angle and mechanical gain.

The *reflex mechanical gain* rose to a peak between 0° and +10° of dorsiflexion beyond neutral, declining steeply thereafter. By contrast, axonally stimulated muscle twitch torque increased serially up to +30° dorsiflexion beyond neutral. For the soleus muscle, the optimal reflex neuro-mechanical angle lies approximately midway between the angle for optimal reflex EMG gain (in mild plantarflexion, at which the largest and strongest motor units appear to be activated) and the optimal muscle mechanical angle (at the extreme of soleus muscle dorsiflexion).

7.4.6.3 Changes in peak reflex force and muscle compliance with age.

Soleus muscle twitch characteristics show a five to eight-fold increase in peak force associated with a ten-fold reduction in compliance in the first two decades of life and an apparent speeding up of twitch time in the first decade.

7.4.6.4 Reflex twitch time, joint angle and age.

The reflex twitch time is more scattered against the joint angle, but increases with dorsiflexion, rising from a mean of 277ms at -25° and 285ms at neutral to 370ms at +35°: a mean difference of 93ms across the joint range, or a 33.5% increase. The slowest twitches

occurred in the youngest children.

7.4.6.5 Evidence for an optimal neuro-mechanical joint angle.

The effects of short-term passive changes in joint angle on stretch-reflex output indicate the influence of peripheral factors on this physiological phenomenon. This explains some of the physiological variability in eliciting and quantitating tendon jerks and velocity-dependent stretch-reflexes within and between subjects as well as the large inter-observer variations in the clinical setting. Much more interesting for motor physiology is the evidence for a functional neuro-mechanical joint range and of optimal neuro-mechanical coupling in mild dorsiflexion beyond neutral: compared to an optimal neural angle of -15° to -10°.

7.5 Discussion.

While there is no doubt that a mechanical tap elicits a stretch-reflex, the hammer blow varies from tap to tap which is why the data was collected as four consecutive tap averages: this was aimed at averaging out the performance of the examiner as well as the subject. If the change in tap-induced reflex EMG output had simply varied with the strength of the tendon taps, there would be no change in reflex EMG gain (ratio of the normalised reflex EMG to normalised tendon tap torque) over the joint range. This indicates a genuine gain in reflex motorunit output in mild plantarflexion beyond neutral, brought about by a change in muscle length which presumably affects the recruitment of the spinal alpha-motorneurones. In addition to the joint angle modulation of the tendon reflex, the joint angle has been shown to modulate the reflex EMG response to sinusoidal stretching in spastic quadriceps and hamstrings muscles in adults (Burke, Andrews and Gillies, 1971) and children (Lin, Brown and Brotherstone, 1994a).

It is clear from the present studies that the optimal joint angles for reflex EMG gain and reflex mechanical gain are out of phase since the reflex mechanical gain achieves its maximum in mild dorsiflexion beyond neutral, the optimal neuro-mechanical angle, which is 20° to 25° more dorsiflexed than the angle of maximum reflex EMG gain (optimal neural angle). When the muscle was stimulated electrically via the posterior tibial nerve motor axons the greatest motor torque arises close to the position of full muscle stretch at +30 of dorsiflexion (a finding which agrees with the work of Sale and colleagues,1982), the optimal mechanical angle. This data suggests that the performance of the muscle cannot be assessed on the basis of the EMG output alone since the mechanical output will vary

according to the joint angle which influences that EMG output differently according to whether the muscle is directly or reflexly excited. This also means that direct axonal stimulation experiments to determine motor output, provide only one picture of the way in which motor output varies with joint angle.

Assuming that the reflex alpha-motorneurone output does indeed vary with the joint angle, is it the change in muscle length or muscle tension which determines reflex excitability? Muscle tension is sensed by the Golgi tendon organs, which are stimulated during *active* muscular contraction but are relatively quiescent in passive changes of muscle length (Houk and Hennemann 1967). This means that the sudden, brief changes in tension produced by the tendon taps themselves are unlikely to be responsible for the changes in reflex excitability, nor are the passively sustained incremental changes in muscle tension produced by the bias torques, leaving either sensory afferents in the ankle joint capsule and soft tissues or, more likely, the muscle spindles themselves as the alternative candidates.

In health, the spinal alpha-motorneurones may be inhibited presynaptically by a number of influences explored by Burke, Andrews and Ashby (1971) in which the H-reflex was inhibited by passive dorsiflexion. This inhibitory influence obtained when the Achilles tendon was stretched (altering its length) but not when it was squeezed (maintaining a constant length but stimulating sensory afferents). When the same investigators selectively blocked large 1a afferent fibres by inducing local ischaemia, the inhibitory effect of stretch persisted indicating that type II (slow conducting) small afferent fibres were responsible for the inhibition of stretch-reflex excitability, such fibres forming the secondary spindle endings which are thought to monitor changes in muscle length (see also Burke and Lance 1973). The effects of calf ischaemia have also been reported by Hultborn and colleagues (1996) who, unlike Burke et al, found that 27 minutes of calf ischaemia abolished the inhibitory effects of previous dorsiflexion on the H-reflex. However, Hultborn and colleagues were looking at the duration of the inhibitory influence of a *previous conditioning dorsiflexion* while studying the H-reflex at a constant muscle length.

The final reflex motor output is thus a summation of the effects of muscle length on neural output, on the output of the contractile elements intrinsic to muscles and the effect of joint angle on the mechanical arrangements of the muscle-fibre-tendon-bony lever complex. This would explain why over the range of a few degrees in dorsiflexion, the reflex peak

torque continues to rise despite the fact that the reflex EMG has started to wane: the mechanical advantage of stretch continues to produce a small increase in peak torque until, with further increments of dorsiflexion, the fall in the number of reflexly recruited alpha-motorneurones is so great that it overrides any mechanical advantage: neuro-mechanical coupling becomes sub-optimal. The optimal angle for neuro-mechanical output is neither the resting angle nor the extreme of dorsiflexion, but midway between the two.

Sale et al (1982), discuss the fact that the calcaneal lever arm (defined as the shortest distance between the line of pull of the Achilles tendon at its insertion and the talo-tibial joint) diminishes by 2/3 in full dorsiflexion from a maximum in plantarflexion. If **Fp** is the intramuscular force in full plantarflexion and **Lp** the length of the lever arm:
the torque **T** generated will be:

$$T=FpXLp. \qquad 7.5.1$$

From this, the relative theoretical intramuscular force,**Fd**, necessary to produce an equivalent torque in full dorsiflexion can be calculated. Assuming the dorsiflexed lever arm to be given by:

$$Ld=2/3Lp \qquad 7.5.2$$

$$T=FdX2/3Lp=FpXLp$$

giving: $$Fd=3/2Fp \qquad 7.5.3$$

This means that for any arbitrary soleus torque in full plantarflexion to be matched by an equal torque in the dorsiflexed (stretched) soleus, the dorsiflexed muscle must actually be capable of generating 1.5 times the plantarflexed intramuscular force. Conversely, the equations would suggest that an arbitrary torque in dorsiflexion can be matched by the plantarflexed (shortened soleus) with only 66% of the intramuscular force-generating capacity of the dorsiflexed muscle, since the lever arm is longer. Therefore, the mechanical leverage helps to offset a reduction in intramuscular force generation of the plantarflexed soleus.

Current texts on basic muscle function (see Mosely, 1992, Rab 1993) continue to stress that the muscle achieves its maximum force at its *resting length* , this being based on the "Blix Curve", which in turn is derived from in vitro experiments in which denervated muscles are held between two fixed points in a dish. This Blix model departs from the data presented here, and that of Marsh et al (1981) and Sale et al (1982), in which the torque generation by the soleus muscle at *resting plantarflexion* is negligible, *resting plantarflexion*

corresponding to the *physiological resting length* of the muscle-tendon complex. The significance of optimal neuro-mechanical coupling for normal physiology can be illustrated by a few examples.

1. In dynamic gait, during the single support phase (stance), the calf muscles act eccentrically to decelerate the tibia as it rotates forwards at the ankle between the first and third rocker phases. Sutherland and colleagues (1980) have demonstrated that during the single support phase,"ankle dorsiflexion was first resisted, then arrested and reversed by plantarflexor muscles acting *eccentrically* , and this eccentric activity occurs between -5° and +10° of the joint range about the neutral angle, followed by a concentric contraction just prior to push-off, which is precisely the joint interval across which neuro-mechanical coupling is maximal in our study.

As stated above, numerous studies of H-reflex modulation during gait have now confirmed the functional importance of this joint range. A practical example of a disturbance of this relationship are the deleterious effects of heel-cord over-lengthening. Under these circumstances the new "optimal" neuro-mechanical angle occurs at a more dorsiflexed position so that the soleus can no longer eccentrically decelerate the shank as it rotates forwards at the ankle, which produces the well-known crouch gait.

2. For innervated muscle, control of the force output is proportional to the number (and size) of the motor units participating in active muscle contraction (the Henneman Size Principle, see Henneman 1974). It has been previously shown that the most forceful, rapid and convenient voluntary alternating movements at the ankle occur close to the neutral joint angle and appear to involve the largest and fastest motor units (Lin, Brown and Walsh 1996, Lin 1997), indicating that certain postures favour the optimal execution of voluntary motor tasks (see section 9).

The amplitude of any alternating movements varying inversely with the square of the frequency of the motion. Accordingly, large joint excursions can only be performed slowly and the fastest movements have small amplitudes (see fig.6.3.3i in section 6 above), oscillating close to the optimal neuro-mechanical angle.

It has been hypothesised, that children may learn to use favourable postures to improve their dexterity in the performance of rapid alternating movements (Lin 1997). As indicated in this study, reflex motor output in resting plantarflexion is weak and brief whereas

in extreme dorsiflexion it is weak and up to 45% slower than that of the resting angle.

Although maximum voluntary contraction appears to override the joint angle modulation of motorneurone output: viz running, (Capaday and Stein 1987), few activities are regularly performed in conditions of maximum voluntary motor drive, and the effect of the joint angle at lower levels of motor activity may be of functional significance in producing economical motor strategies. It is likely that the spinal alpha-motorneurone pool undergoes presynaptic modulation during physiological movements, being relatively refractory to reflex recruitment at the extremes of the joint range, except at the expense of greater descending motor drive. at the extremes of the joint range, only the small, weak, low-threshold motor units are recruitable.

3. Involuntary alternating movements, such as ankle clonus are likely to be influenced by the joint angle: the extremes of the joint angle being refractory to clonus because of low neuro-mechanical output, the strongest beats occurring just beyond neutral, becoming slower and weaker with progressive dorsiflexion as the reflex EMG and reflex torque dwindle and the muscular twitches get slower. The clonic phenomena are likely to change after peripheral interventions such as tendon lengthening or plaster immobilisation at an increased muscle length, which would shift the excitability curve to the right as well as have an effect on the contractile properties of the muscle ie alter fibre type composition. These predictions have been explored in section 8.

7.6.1 Conclusions.

i. The soleus muscle reflex excitability is maximal at -10° of plantarflexion beyond neutral.

ii. The soleus muscle reflex mechanical twitch is maximal at 0° to +10° beyond neutral at a time when the reflex EMG response has begun to wane.

iii. The soleus muscle reflex twitch time increases by 33% over the resting plantarflexed value.

iv. The findings, which have been briefly reported for healthy adults (Lin, Brown and Walsh, 1996b) strongly support the concept of a continuum of reflex excitability and of a functional joint range in which an optimal soleus muscle "neuro-mechanical" angle between +/-10° on either side of neutral can be specified.

v This optimal muscle length is different to the "resting length".

vi The findings confirm the role played by peripheral factors in the subtle regulation of apparently "centrally-defined" motor phenomena and provide predictive information on the likely alterations in behaviour of the the soleus muscle in health and when held at pathological joint angles (postures).

These findings may be of relevance to current orthopaedic practice, sports medicine, rehabilitation and associated physical therapies, in focussing attention on how this reflex modulation might influence motor function in health, injury and disease states. Interventions may produce a new motor picture which can be partly predicted from this joint angle model.

8. Soleus Reflex Twitch Characteristics in Childhood Hemiplegia

8.1 Background.

Most treatments used in cerebral palsy involve some form of physical manipulation of the peripheral motor apparatus comprising the bones (osteotomies, derotations), the muscles and non-contractile soft tissues (stretches, exercises, orthotics, serial plastering, tendon releases, lengthenings and transfers, electrical stimulation) the neuromuscular junction (botulinum toxin A injections), and the peripheral nerves (alcohol and phenol nerve blocks, selective and unselective dorsal rhizotomy). Such treatments are aimed at increasing function by preventing or relieving deformity, reducing "phasic spasticity" (unwanted velocity-dependent motor activity), "tonic spasticity" or "dystonia". All these treatments, including the centrally and spinally acting tone-relieving drugs such as the benzodiazepines and baclofen, assume that the neurological condition is non-progressive and in particular that the physical expression of the motor disorder is invariant.

The pathophysiological changes in the effector organs (muscles and tendons) are seldom if ever considered beyond a clinical assessment of the presence or absence of contracture. The muscles are viewed as passive instruments controlled by disordered brain and spinal mechanisms.

Identifying specific mechanisms and changes brings us closer to the development of appropriate treatments aimed at preventing or reversing the consequences of brain damage. The purpose of these investigations was to explore the changes in the contractile elements of muscle and to determine how, if at all, these changes contribute to the symptoms and signs in cerebral palsy. The short-term influence of varying the joint angle by passive stretching of the soleus muscle on EMG discharge and twitch characteristics following the ankle jerk in healthy adults and children has been studied in section 7 and published in part elsewhere (Lin, Brown and Walsh, 1994, 1996a and b;Lin 1997).

In the present studies, the soleus reflex twitch characteristics of children with hemiparetic cerebral palsy are investigated. The differences between those hemiparetic limbs exhibiting clinical clonus are compared with "non-clonic" hemiparetic limbs. Follow-up data on the effects of heel-cord lengthening and of muscle immobilisation at neutral on the soleus muscle in the congenital hemiparetic limb is presented in the next section which provides a further clue to the mechanisms of clonus and how its expression may be modified

by peripheral interventions.

These studies indicate that modifying the "engine" of the motor system (the muscles) modifies the expression of the motor neurology, with particular reference to reflex excitability and the tendency to exhibit clonus. The results suggest, that just as the ability to perform voluntary alternating movements is bounded on the one hand by the laws of physics governing mechanical oscillation and on the other by the force-generating capacity and speed of muscles (see below), so the frequency of clonus appears to depend on the twitch characteristics of muscles, which in turn may be modified by a variety in internal and external stimuli.

An appreciation of the influence of disuse, or on the contrary, muscle activity on such processes throws light on the natural history of cerebral palsy and the possible consequences of treatment. Paradoxically the phenomenon of clonus may be used as a sensitive index of the effect of peripheral treatment on the muscles and the way the muscles interact with the central nervous system.

8.2 Methods.

The apparatus and experimental arrangements for measuring the virtual isometric reflex twitches in the soleus muscle has been described in detail in section 7, above and the data collected in the same way as for the adult and paediatric controls. The study was conducted with full approval of the Lothian Heath Board Paediatric and Reproductive Medicine Ethics committee.

The subjects include a convenience sample of 22 healthy control children aged 3.9-13.6 years, details of which have been previously reported (Lin, Brown and Walsh, 1994, 1996b, 1997a, b). There were 18 hemiparetic children studied aged 3-14.9 years. Limbs were classed according to whether they were controls, nonparetic (the 'uninvolved' side of a hemiparetic child), hemiparetic without evidence of clinical clonus (non-clonic) or hemiparetic with evidence of clinical clonus (hemiclonic). Observations for all subjects included:

8.2.1. Anthropometry.

Age (years); sex; weight (Kg); stadiometer height (cm); fibula length (cm) using an inextensible tape measure; foot length (cm) using callipers; maximum calf diameter (cm) with callipers and maximum calf circumference (cm) using an inextensible tape measure.

8.2.2. Biomechanical variables.

Measurements were made as previously described (Lin, Brown and Walsh 1994,1996b) with the subjects lying comfortably on the left side, to eliminate the influence of gravity, with the hip and knee flexed to 90° to eliminate the effects of the gastrocnemius muscle (Silverskjöld, 1923) which crosses the knee as well as the ankle joint, and is relaxed when the knee is flexed. For details of the high inertia beam, force plate and printed torque motor see Walsh et al 1993 and section 4.3.1 above.

Using this apparatus, measurements included the resting angle (°) at the ankle; the applied bias torque (Nm) to produce passive soleus muscle stretch; the bias angle (°) produced by the bias torque, the relative angular displacement (°) which is the angular interval from the resting angle to the bias angle; the soleus muscle compliance (°/Nm), which is derived from the angular displacement divided by the bias torque and is expressed in degrees of angular displacement (°) per unit of bias torque (Nm).

8.2.3. Neurophysiological variables.

Reflex soleus muscle twitches were elicited by blows to the Achilles tendon as described in section 7.3.1-5 above. Parents were present throughout for all children studied. As for previous studies, measurements included the tendon jerk reflex EMG (mV) and the mechanical events included the reflex twitch: peak force (N), 1/2 contraction time (ms), 1/2 relaxation time (ms), total twitch time (ms) and the reflex twitch frequency (Hz) which is the inverse of the reflex twitch time. The 1/2 contraction and 1/2 relaxation times correspond to the almost linear phases of contraction and relaxation which occur between 25% and 75% of the peak force ie the interquartile temporal characteristics. For definition of these neurophysiological variables see fig. 7.3.8.2, above.

In some individuals no reflex was obtainable until the soleus muscle had been stretched by applying a dorsiflexing torque to the beam with the motor. A typical study lasted an hour for two limbs, with time to rest briefly between joint increments and between measurements of the right and left legs.

8.2 4 Data analysis.

All results were analysed off-line. The presentation of the results relates to:

8.2.3.1. Comparative data between groups.

The data for control, nonparetic, hemiparetic limbs without clinical clonus (HP no clonus) and hemiparetic limbs with clinical clonus (HP clonus) are compared for all the

measured variables. Measurements of each variable for each group are expressed in terms of the mean and 99.99% confidence intervals (99.99% CI). These groups were compared using an analysis of variation (ANOVA, Statview + graphics) technique.

8.2.4.2. Follow-up measurements.

This includes sequential data obtained from four subjects, the intervals between observations ranging from 2 weeks to 1 year. Three children were followed up before and after casting and one child, who was first studied 6 months after his second heelcord operation was restudied a year later, i.e 18 months after surgery. Where necessary, the physiological traces and analysed data are illustrated.

8.2.5 Clinical background for the hemiplegic children.

One of the hemiparetic children was not receiving any specific treatment at the time of the study, seven cases had recourse to physical therapy in varying amounts and in varying combinations and this also included one child who was using night splints. Eight cases were using ankle foot orthoses. Two cases had previously undergone heel-cord surgery in early childhood and more recently within 2 years of the study in one case, and within 6 months in the case of the other. A larger number of children had undergone serial casting at some time in the past.

8.3 Results.

8.3.1.Anthropometry.

The anthropometric details for each clinical group are given in table 8.3. which shows that the 22 control and 18 hemiparetic children did not differ in age, height, weight, fibula length, maximum calf diameter and circumference or foot length. The confidence intervals for the clonus limbs are very wide because there were only four hemiparetic limbs with clonus.

8.3.2 Biophysical parameters.

The biophysical variables for each limb grouping are given in table 8.3.which gives the means and 99.99% confidence intervals.

Limb Status		Control: 42 limbs 22 cases			Nonparetic: 18 limbs			HP no clonus: 14 limbs			HP clonus: 4 limbs		
		mean	99.99% CI lower	upper	mean	99.99% CI lower	upper	mean	99.99% CI lower	upper	mean	99.99% CI lower	upper
Age	yrs	7.65	6.0	9.2	7.60	4.8	10.3	6.74	4.0	9.45	10.08	-3.8	24.0
Height	cm	125.86	115.41	136.3	123.5	106.9	140.2	118.7	102.1	135.3	138.7	60.5	216.9
Weight	Kg	26.67	21.24	32.1	26.4	17.28	35.46	24.1	15.13	33.24	32.41	-17.96	82.79
Fibula length	cm	28.1	25.3	31.0	27.18	19.5	34.86	25.5	16.87	34.12	30.5	-50.73	111.73
Calf diameter	cm	7.3	6.7	7.9	7.3	5.9	8.7	6.5	5.2	7.8	6.92	-10.7	24.5
Calf circumference	cm	24.5	21.8	27.1	25.0	20.2	29.7	22.9	18.1	27.7	24.2	-44.0	92.5
Foot length	cm	20.3	17.9	22.7	18.9	15.1	22.6	18.0	13.8	22.2	20.3	-25.2	65.8
Resting Angle	°	-13.5	-17.8	-9.3	-15.8	-22.0	-9.5	-22.42	-34.35	-10.5	-14.25	-170.72	142.2
Bias Torque	Nm	0.77	0.63	0.9	0.69	0.55	0.84	0.91	0.70	1.13	0.85	0.50	1.20
Ankle Bias Angle	°	4.5	1.7	7.4	2.35	-0.74	5.44	-2.74	-6.0	0.59	3.06	-4.14	10.27
Ankle Joint Displacement	°	18.6	15.8	21.4	17.62	14.6	20.63	17.63	14.48	20.78	15.7	10.67	20.73
Compliance	°/Nm	28.8	23.7	34.0	28.53	24.42	34.65	25.18	19.27	31.08	25.51	12.17	38.84
Ankle Reflex EMG	mV	1.0	0.79	1.2	**2.85**	2.1	3.6	**3.21**	2.43	3.99	**4.47**	3.17	5.78
Reflex Twitch Force	N	5.88	4.9	6.86	9.6	7.74	11.46	12.05	9.5	14.8	11.27	7.15	15.48
1/2 Contraction Time	ms	37.1	34.2	40.0	35.28	32.38	38.19	**43.23**	39.9	46.49	31.14	25.58	36.7
1/2 Relaxation Time	ms	69.2	62.3	76.1	62.46	54.1	70.82	**76.08**	63.01	89.15	**44.06**	33.2	54.93
Total Twitch Time	ms	297	285.4	308.6	295.92	278	313.8	**330.56**	307.58	353.54	**235.81**	188.97	282.66
Twitch Frequency	Hz	3.46	3.33	3.58	3.5	3.32	3.68	**3.17**	2.96	3.38	**4.77**	3.53	6.0

Table 8.3. Anthropometric, biomechanical and neurophysiological variables.
Data from 42 control limbs, 18 nonparetic limbs, 14 hemiparetic limbs without clonus and 4 hemiparetic limbs with clonus.

8.3.2.1. a. Resting angles of plantarflexion and b. bias torques.

Table 8.3 and figures 8.3.2.1a and b illustrate the mean and 99.99% CI for the resting angle and dorsiflexing torques respectively. There were no differences between the mean resting angles and applied bias dorsiflexing torques respectively of the control (-13.5° and 0.77 Nm), nonparetic (-15.8° and 0.75 Nm) and hemiparetic limbs with clonus (-14.2° and 0.85 Nm). However the mean resting angles and applied dorsiflexing torques of the hemiparetic limbs without clonus were significantly different from those of control limbs, these displaying a greater degree of resting plantarflexion (-22.4°) and requiring larger dorsiflexing torques (1.033Nm), $p<0.0001$ respectively.

8.3.2.1c Bias angles, d. angular displacement and e. calf compliance.

The mean bias angles of the 14 hemiparetic limbs without clonus (fig 8.3.2.1c) were consistently lower at a mean of -2.7° of plantarflexion compared with+3° beyond neutral for the 4 hemiparetic limbs with clonus, + 2.3° beyond neutral for the 18 nonparetic limbs and +4.5° beyond neutral for the 42 control limbs. The mean bias angle of the hemiparetic limbs without clonus was significantly more plantarflexed than that of the nonparetic or control limbs ($p< 0.001$). However, taken as a group, all 80 limbs were subjected to similar angular displacements at the ankle averaging 14-21° (fig. 8.3.2.1d) and this produced statistically similar average compliance values of 21-28°/Nm for all groups (fig.8.3.2.1e). Nevertheless, for some cases, there was considerable within individual variation between nonparetic and hemiparetic limbs. These results are also shown in table 8.3.

8.3.3 a-d Neurophysiological parameters.

The means and 99.99% CI are summarised in table 3.8 and figure 8.3.3 a-f.

8.3.3 a Reflex EMG.

The highest peak-peak reflex EMG amplitudes were obtained in the hemiparetic limbs with clonus (fig. 8.3.3a) (4.79mV) which differed significantly from the means of the control (1mV), nonparetic (2.85mV) and nonclonic limbs (3.21 mV) respectively. It should be noted that the reflex EMG of the nonparetic and hemiparetic limbs without clonus also differed significantly from that of controls ($p<0.0001$), but the EMG discharges of nonparetic limbs were similar to those of hemiparetic limbs without clonus. These should be interpreted with caution since the EMG data are not normalised within or between limbs or subjects, but the surface electrode positioning was standardised.

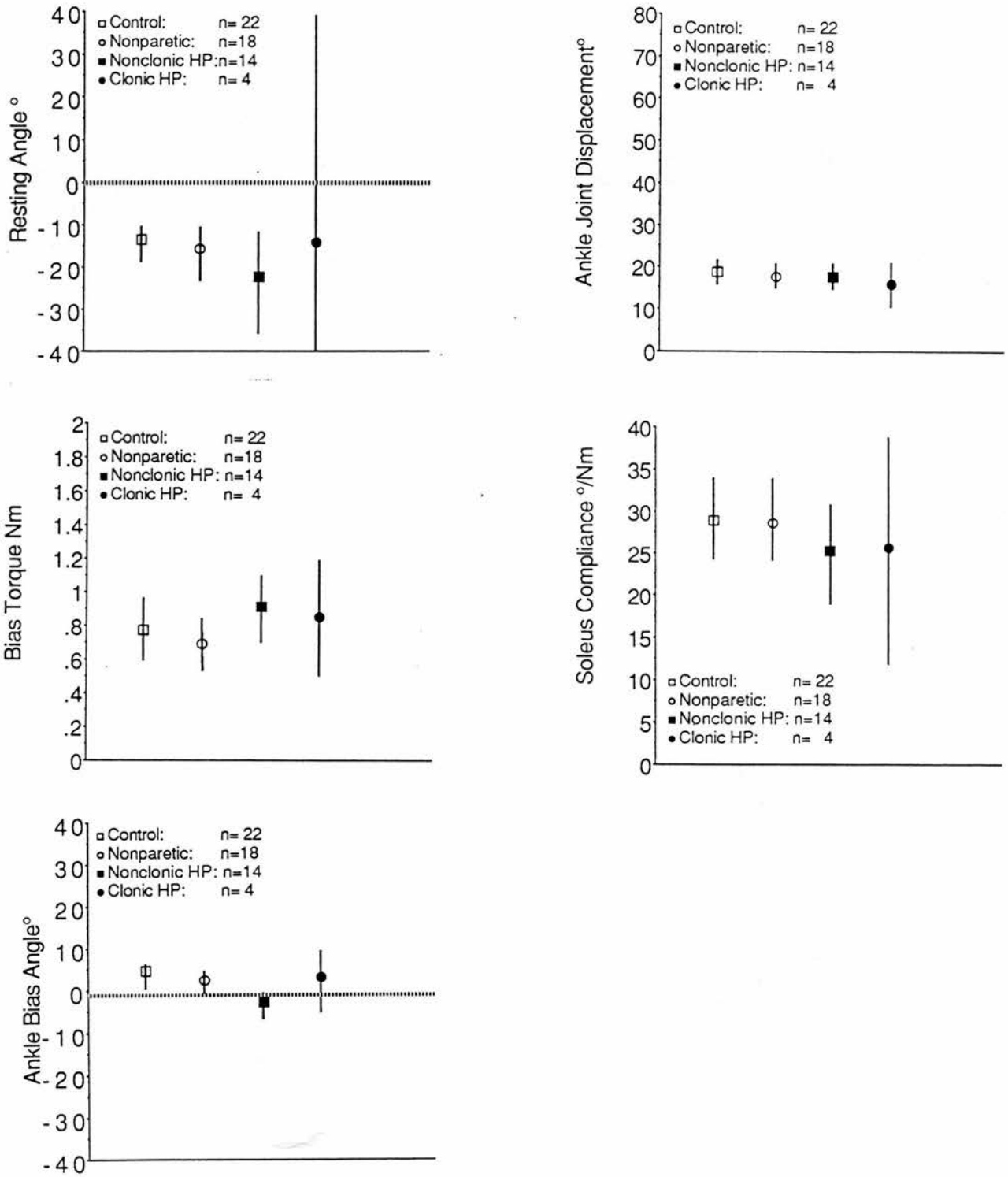


Figure 8.3.2.1a-e Biomechanical Variables.
Means and 99.99% upper and lower CI for **a**. resting ankle joint angle (°), **b.** applied dorsiflexing torque (Nm), **c.** bias angle (°), **d.** angular displacement (°), **e.** compliance (°/Nm). Control limbs =22, nonparetic limbs =18, nonclonic limbs=14, clonic limbs=4

8.3.3.b Reflex twitch force.

The control limbs (fig. 8.3.3b) had the weakest mean reflex twitch forces (5.88N) compared with the other groups ($p<0.0001$). The highest mean reflex twitch forces occurred in the nonclonic (12.05N) followed by the clonic (11.27N) and the nonparetic (9.6N) limbs.

8.3.3.c Reflex half contraction time (1/2 CT).

The slowest mean 1/2 CT (fig. 8.3.3c) were found in the nonclonic limbs (42 ms) which were similar to values for the control limbs (37 ms) but differed from the mean for the nonparetic limbs (32 ms) and the clonic limbs (31 ms) which had the fastest mean 1/2 CT of all ($p<0.0001$). It should be emphasised that the major contribution to the reduced 1/2 CT came from the limb which had undergone heel-cord lengthening six months previously (see follow-up' studies below).

8.3.3.d Reflex half relaxation time (1/2RT).

The nonclonic limbs (fig. 8.3.3d) had the slowest mean 1/2RT (78 ms) which resembled values obtained for the control (69 ms) and nonparetic (64 ms) limbs. The clonic limbs had the fastest mean 1/2 RT of 41 ms which differed significantly from the control, nonparetic and nonclonic limbs ($p<0.0001$). Once again, the most striking contribution to this reduced 1/2 RT came from the case with heel-cord lengthening within the last six months (see 'follow-up' below).

8.3.3.e Reflex twitch time.

The slowest total reflex twitch times (fig.8.3.3e) belonged to nonclonic limbs (330 ms) although these total twitch times matched values for nonparetic (295 ms) and control limbs (297 ms). The clonic limbs had markedly faster mean twitch times of 235 ms compared with other groups ($p<0.001$) and were almost 100ms faster than the non-clonic limbs. All muscles exhibited longer twitch times with increasing dorsiflexion and the regression equations for the curves representing the scatter of reflex twitch times against joint angle give values for the mean twitch times at neutral of 336 ms for the nonclonic, 292 ms for the nonparetic and 285 ms for the control limbs. These twitch times closely match the above quoted mean values which were calculated across the joint range. Dorsiflexion beyond neutral (0°) exerts a profound effect on all limbs, adding upwards of 100ms to the total reflex twitch time near maximum dorsiflexion.

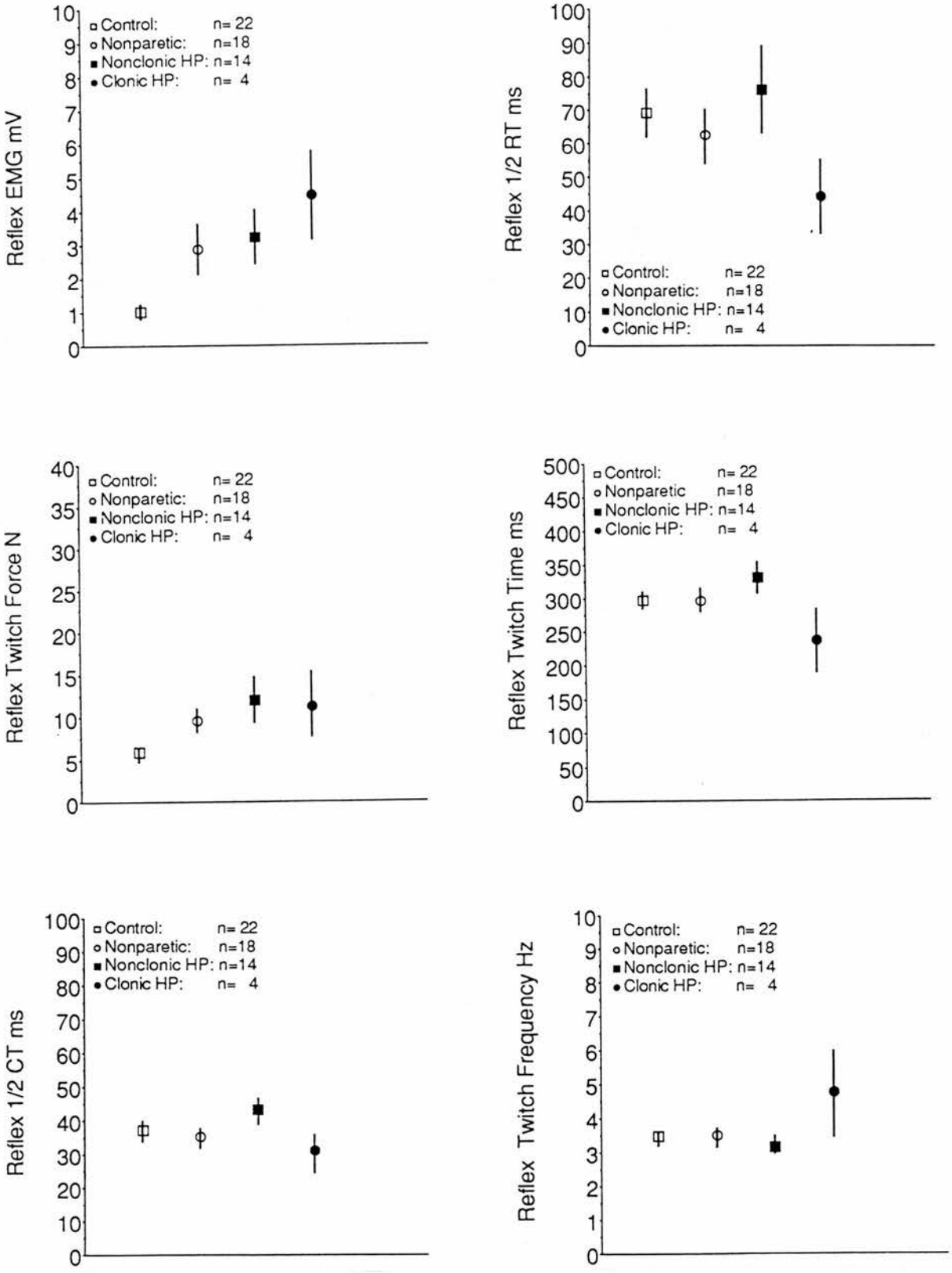


Figure 8.3.3 a-e. Neurophysiological Variables. Means and 99.99% upper and lower CI for control, nonparetic, nonclonic and clonic limbs of hemiparetic children. **a.** reflex EMG (mV), **b.** reflex twitch force (N), **c.** 1/2 contraction time (ms), **d.** 1/2 relaxation time (ms), **e.** Total twitch time (ms) and **f.** reflex twitch frequency (Hz). Clonic limbs have fastest twitches.

8.3.3.f Reflex twitch frequency.

The maximum reflex twitch frequency (fig. 8.3.3f) represents the theoretical number of consecutive reflex twitches which can be added end to end per second or inverse twitch time. Accordingly, the clonic limbs had the highest mean maximum twitch frequencies of 4.8Hz, which clearly differed from nonparetic and control twitch frequencies of 3.4 Hz and the non-clonic frequencies of 3.2 Hz ($p<0.0001$).

8.3.4. Reflex twitch time in clonic limbs

No single regression equation fits the clonic limbs as a group (fig. 8.3.4), but each individual case within this group clearly obeys the same principle indicated in fig. 8.3.3e, of increased twitch time with dorsiflexion beyond neutral, following what appear to be a concentric family of twitch-time curves for each of the four clonic subjects. The extremely brief twitches for the clonic group are clearly heavily influenced by values obtained in the 6 year old boy who had undergone heelcord surgery six months previously and whose muscles had exceptionally fast contraction and relaxation times. The reflex EMG and twitch characteristics of this case are shown in figure 8.4.1 at different passive joint angles (soleus stretches). Nevertheless, two other limbs exhibiting clonus also had twitch times at the lower end of the range for the group as a whole (fig. 8.3.4).

8.3.5 Reflex twitch frequency, joint angle and clinical clonus.

The functional significance of the changes in twitch frequency with joint angle are illustrated in figures 8.3.5 a-d, which demonstrate the fastest twitch frequencies of some 4-5 Hz in -20° to -30° of plantarflexion compared with 2-3Hz in 20° to 30° of dorsiflexion beyond neutral. This clearly reflects changes in the functional twitch characteristics of the muscles with increasing muscle length. The fastest twitch frequencies for all groups of limbs occur in plantarflexion. Accordingly, the fastest clinical clonus was observed at angles of relative plantar flexion in three cases and the clonus twitch frequency was observed to slow with gradual increases in dorsiflexion, the clinical clonus being abolished or unobtainable beyond neutral. In each case, there was a joint angle at which sustained clonus could be elicited, further dorsiflexion beyond which, clonus faded out.

The fourth of the clonic limbs exhibited an extreme departure from the normal range with twitch frequencies achieving maximum values of 11Hz at +6° falling to a minimum of 6Hz at +20° of dorsiflexion (figs. 8.3.5d and 8.4.1). The joint angles for sustained clonus in this

case were wholly in dorsiflexion beyond neutral, but passive stretching steeply reduced the twitch frequency by almost half and also had an inhibitory effect on the number of beats of clonus, inducing" fade" from about +14° of dorsiflexion onwards as shown in (figures 8.3.6).

This data, based on the reflex twitch profile of the muscles, indicates that the fastest clonic movements will occur in the muscles with the fastest twitch profiles. The joint angle, twitch frequency and ability to elicit and sustain clonus in a hemiparetic limb appear to be closely related phenomena.

An additional factor contributing to the frequency of oscillatory motion is the twitch force which can be shown to vary with the square of the frequency of oscillation (see section 9 and Walsh, 1992; Lin, Brown and Walsh, 1996a and Lin 1997). According to this law, a doubling in clonus frequency would require a four-fold increase in muscle force and a trebling of the clonus frequency a nine-fold increase in twitch force, so that the fastest clonus beats must occur at joint angles permitting the fastest and strongest reflex contractions: near the neutral angle being the optimal joint position for obtaining sustained clonus, unless the the muscle twitch joint angle characteristics are upset by pathology or intervention (s). This issue of the optimal joint angle, muscle twitch force and frequency is clarified by follow-up studies in four children.

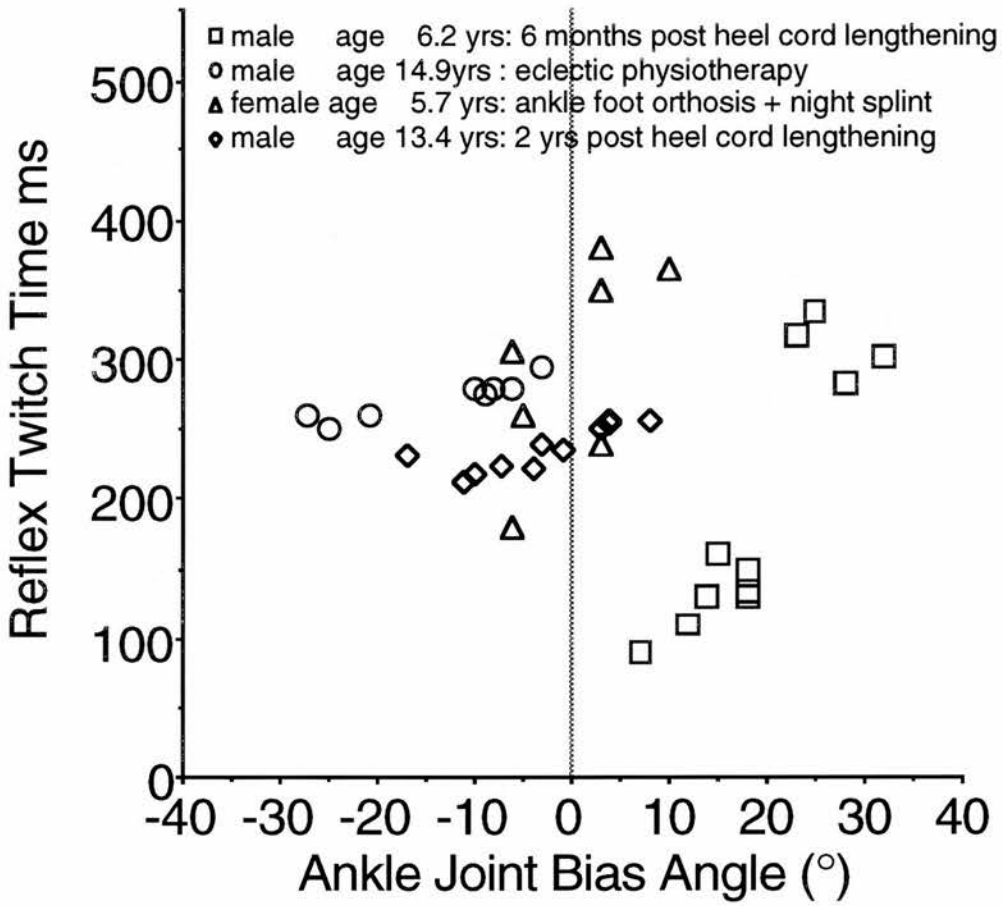
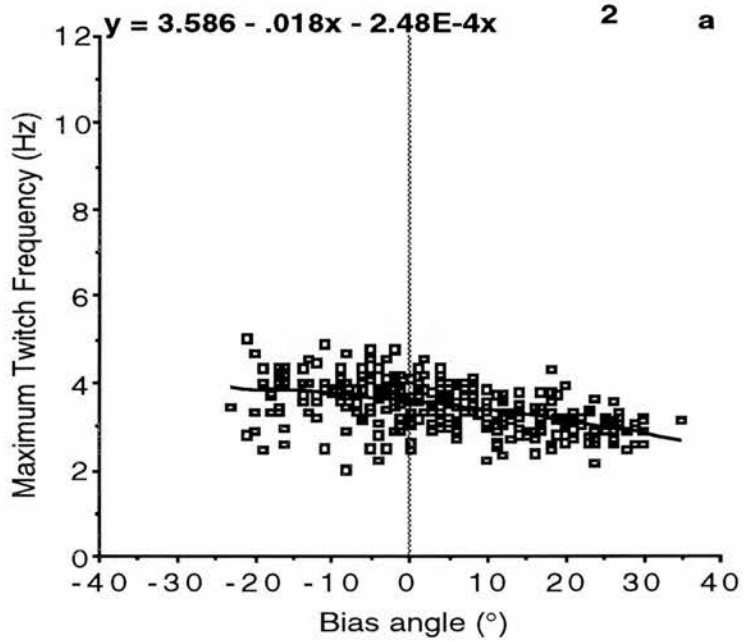


Figure 8.3.4 Clonic limb twitch times against joint angle.

Clonic limb twitch time (ms) against joint angle (°). The clonic limbs form a family of four distinct twitch-time curves across the joint range. Overall, faster twitch times occur in those muscles which have undergone heelcord lengthening followed by orthotics and physical therapy.

The fastest values were obtained in a 6 year old boy who had undergone heelcord surgery six months previously and whose muscles had exceptionally fast contraction and relaxation times (square symbols)



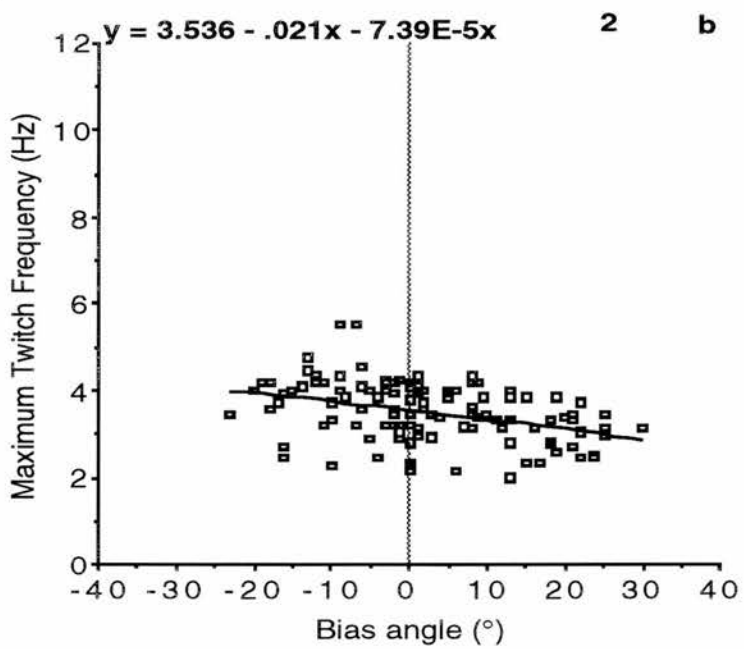


Figure 8.3.5 a-b Decline in reflex twitch frequencies with dorsiflexion.
Similar distribution of reflex twitch frequencies (Hz) across the joint range for:
a. control. and b. nonparetic limbs. In one of the nonparetic limbs, no reflex was elicitable.

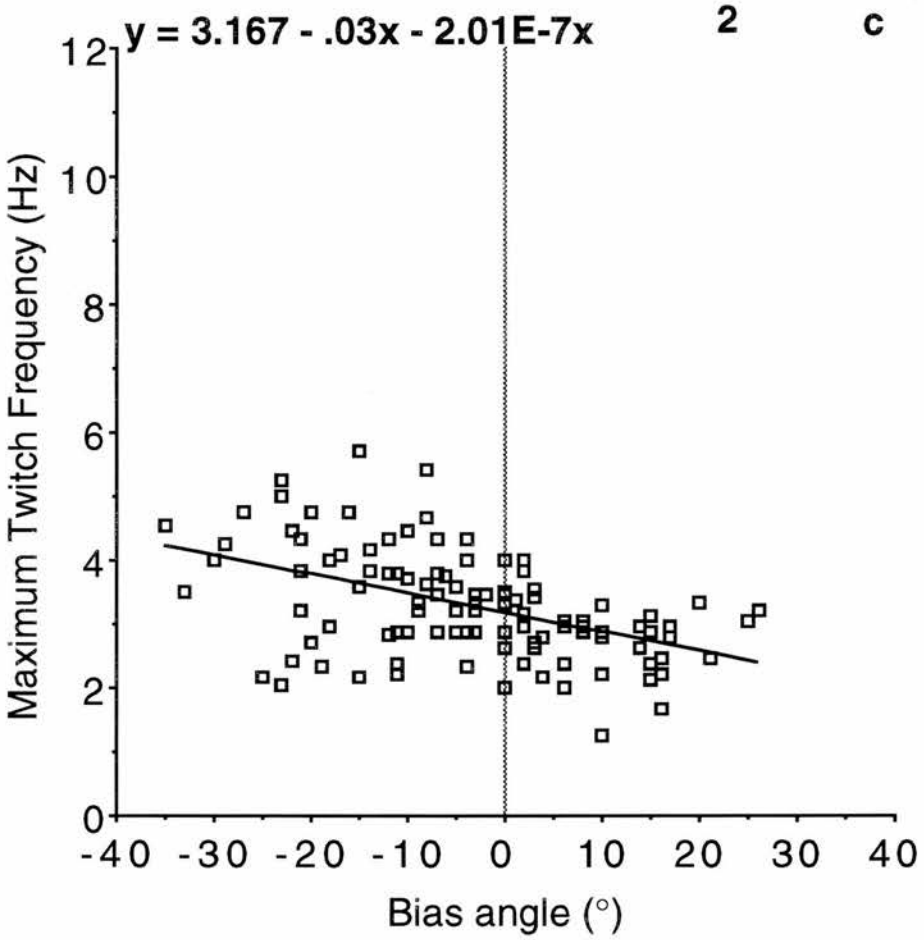


Figure 8.3.5 c Decline in reflex twitch frequencies with dorsiflexion.
c.nonclonic limbs. The nonclonic limbs were the most plantarflexed for the group as a whole. For clonic limbs, see 8.3.5.d (below)

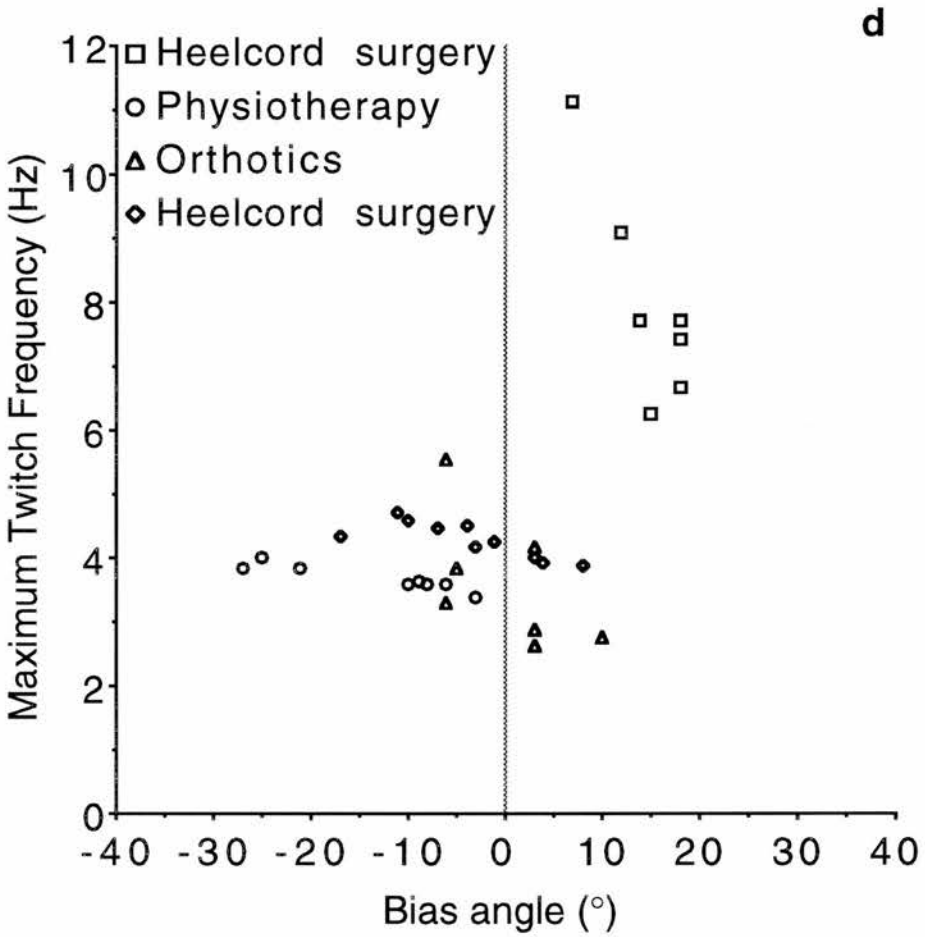


Figure 8.3.5 d Decline in reflex twitch frequencies with dorsiflexion.
d. clonic limbs.
The clonic limbs are capable of the fastest repetitive twitches.
Clonic limb twitch frequency also declines with dorsiflexion.
For control, nonparetic and nonclonic limbs see 8.3.5 a-d (above)

8.4 Follow-up measurements after interventions to relieve equinus.

Four children were followed up to determine the possible effects of a specific intervention to relieve equinus of the hemiparetic limb. One child was studied following heelcord lengthening and three children before and after plaster immobilisation of the ankle joint at the neutral angle.

8.4.1. Soleus reflex twitch characteristics after surgical heelcord lengthening

It is clear from the above group studies comparing the reflex twitch characteristics of control, nonparetic, nonclonic and hemiclonic limbs that there appears to be a significant interaction of:

i. muscle length-joint angle

ii muscle twitch phenotype (ie strong and slow or weak and fast)

iii reflex excitability

The interaction of these three phenomena are demonstrated by the case of a six year old boy with a congenital right hemiparesis who had been walking from the age of 18 months and had undergone heel-cord lengthening at the ages of 2.5 and 5.8 years, the last lengthening being six months prior to enrolment to the study for the correction of an equinus pattern of walking (toe striking). At the first clinical evaluation, the hemiparetic limb was thin, the ankle jerk brisk, the plantar response extensor and sustained clonus was clinically elicited. The foot came to rest at the neutral position (right angle to the tibia) instead of in slight plantarflexion and could be fully dorsiflexed without encountering even the usual normal resistance. One year later, the clinical ankle jerk of the hemiparetic limb appeared less brisk and ankle clonus could no longer be sustained. Figure 8.4.1 represents follow-up recordings from the nonparetic and hemiparetic soleus. The first and second horizontal sequences of figure 8.4.1 show the EMG and force recordings following single tendon taps to the nonparetic limb successively at increments of +6°, +13° and +21° of dorsiflexion beyond neutral. The near synchronous EMG discharge is followed by a single soleus twitch at each angle. Note the diminution in reflex EMG discharge at + 21° of dorsiflexion and the correspondingly weaker and slower muscle twitch at this angle. The third (EMG) and fourth (force) horizontal sequences show the effects of single taps to the hemiparetic Achilles tendon 6 months following heel-cord lengthening at successive joint increments of +7°, +14° and +18° of dorsiflexion.

Legend to figure 8.4.1 Clonus and the joint angle after heel-cord lengthening.

see overleaf

Top two sequences: nonparetic EMG and force (6 months after heelcord lengthening of the hemiparetic limb) at +6°, +13° and +21° of dorsiflexion beyond neutral.
Middle two sequences: hemiparetic EMG and force 6 months after heelcord lengthening at +7°, +14° and +18° of dorsiflexion beyond neutral:
Lowest two sequences: hemiparetic EMG and twitch force eighteen months after heelcord lengthening at +9°, +15°, +18° and +35°. Note that the clonus frequency declines with passive dorsiflexion both at 6 and 18 months post surgery. The tenotomised muscle becomes slower after 18 months post surgery and the clonus frequency is correspondingly is halved. For further details, see text.
Vertical scale: EMG = 1mV, Force = 6.37N. Horizontal scale = 100ms.

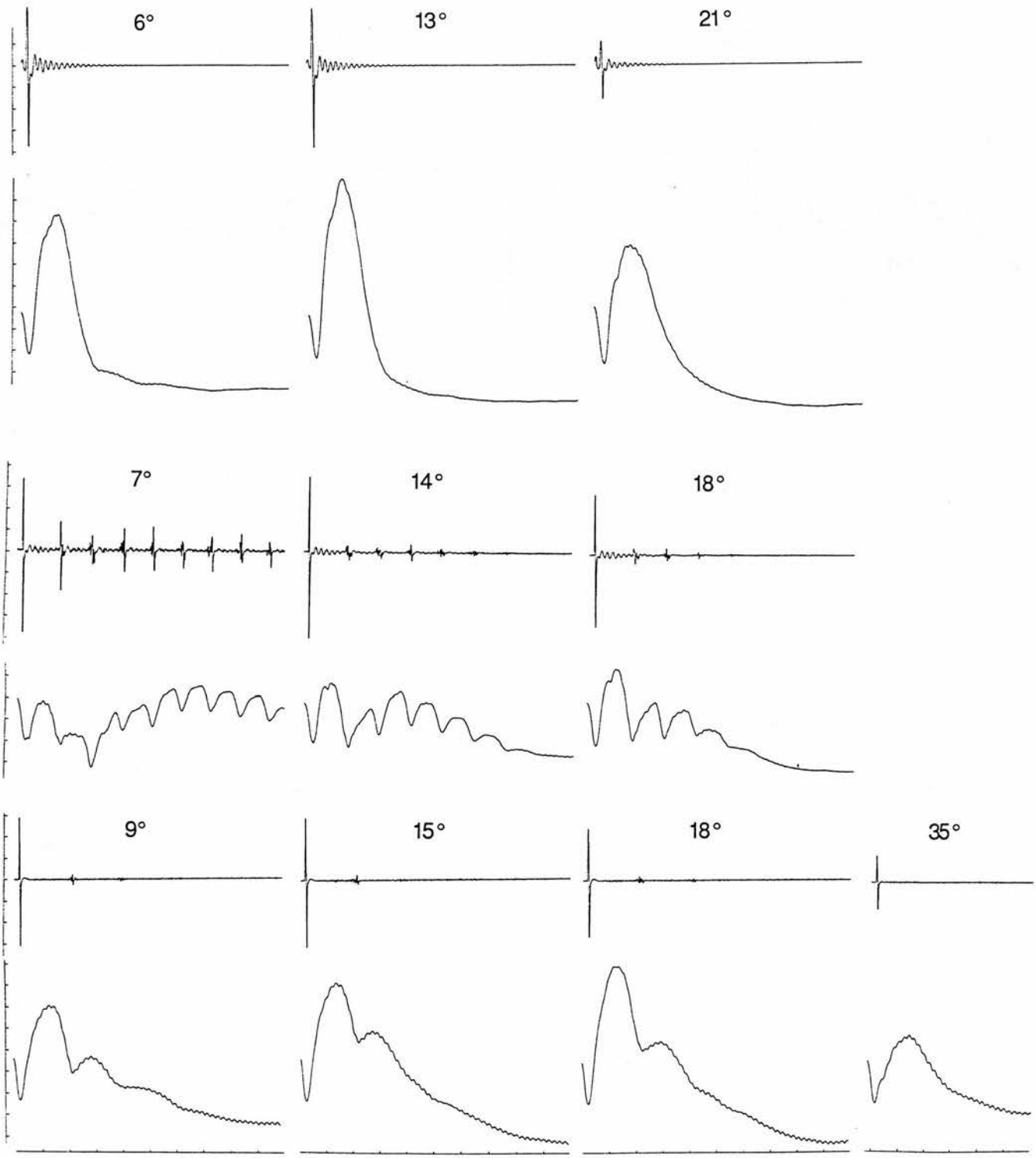


Figure 8.4.1 Clonus and the joint angle after heel-cord lengthening.
For explanation, see legend and text, above and below.

It had not been possible to obtain a tendon reflex in plantarflexion or at neutral. The first near-synchronus EMG discharge after the tendon tap in the hemiparetic limb occurs at +7° of dorsiflexion beyond neutral. This is followed by an extremely rapid and brief soleus muscle contraction and relaxation, the end of which coincides with a second, less synchronous (broader) and polyphasic EMG discharge representing the first clonus beat, followed by a regular and sustained clonus sequence at a frequency of 9 Hz with partial tetanisation of the soleus muscle contraction. The ankle jerk at +14° of dorsiflexion produces a stronger and slower initial contraction and only six clonus beats are now elicited. The reflex EMG discharges giving rise to the clonus beats at +14° and + 18° of dorsiflexion are of lower amplitude and are less phasic than the discharges at +7°. The reflex mechanical twitch increases in force and duration at +14° and +18° of dorsiflexion respectively while the clonus frequency falls to 8.3Hz at +14° and 7.6Hz at +18°.

The reflex excitability is still modulated by passive stretch to the tenotomised muscle. The clonus EMG discharges correspond to the troughs in twitch force ie to when twitch force or intramuscular accelerations are zero or near zero (allowing for the fact that tetanisation alters the baseline) at a time when intramuscular relaxation velocity would be expected to be maximum. In engineering terms, the clonus EMG shows a 90° 'phase lag' with the force trace, indicating that it is velocity-dependent. These findings demonstrate that there is an optimal joint angle for eliciting ankle clonus: in this case, the clonus frequency depends on the excitability of the neuromuscular complex dictated by the joint angle which influences the intraspinal influences on the motorneurone pool and the intrinsic muscle twitch characteristics. Passive dorsiflexion to+14° and +18° is capable of slowing the rate of clonus.

This led to the prediction that a further intrinsic slowing of the twitch characteristics of the soleus muscle would reduce the clonus frequency. Conversely, should the muscle undergo a faster-twitch transformation for any reason, the clonus frequency would rise. The peripheral twitch characteristics of the muscle is apparently crucial for the clinical expression of clonus.

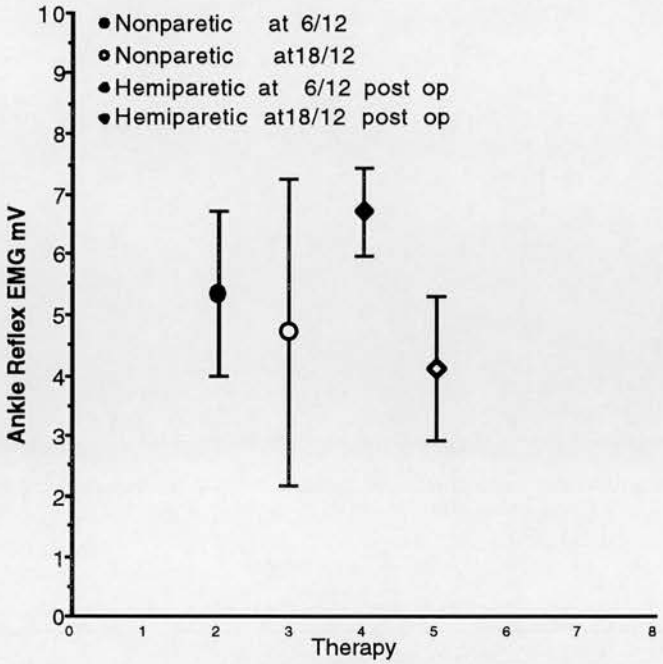
This hypothesis was tested by remeasuring the hemiparetic limb one year later. During the intervening year, the subject had been fully ambulant and had taken active part in school soccer and physical education. The fifth (EMG) and sixth (force) horizontal sequences represent measurements of the hemiparetic soleus made one year after the first recording or

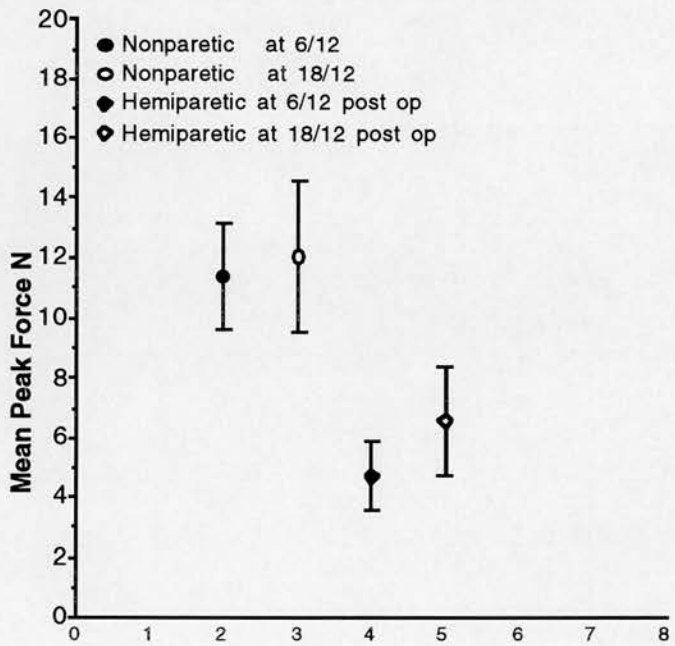
eighteen months following heelcord lengthening. The first reflex muscle twitch at 9° of dorsiflexion is more than twice as strong and twice as long-lasting as that measured one year before: it now resembles the twitch profile of the nonparetic limb while still remaining weaker in absolute terms. The timing of the first clonus EMG discharge now occurs some 200ms after the first reflex EMG discharge instead of 150ms the year before and the interval between successive EMG clonus discharges is 200ms compared with 110ms the previous year. At the neutral joint angle, only two low amplitude EMG bursts of clonus with a frequency of 5.5Hz can now be recorded, falling to 5.2Hz at +9° compared to 8Hz at an equivalent joint angle the previous year (see the relative amplitudes of the clonus to tendon tap EMG discharges for a comparison of relative scale a year apart) and each EMG burst lasts almost twice as long, ie is even more dispersed than before. When the soleus is now stretched successively to +15° and then +18° of dorsiflexion, the strength of the first reflex twitch continues to increase while the clonus frequency falls to 5Hz and 4.5Hz respectively, falling to 4Hz at +26° until at +35° of passive dorsiflexion, both the reflex EMG and twitch force are reduced. At this angle, the total twitch time is markedly slow and no clonus beats are now elicited.

It can be seen that the twitch profile at all angles more closely resembles the reflex twitches in the nonparetic limb, the hemiparetic muscle having undergone a fast to slow transformation over the course of the year, though still retaining a faster twitch profile than the nonparetic muscle. Not only is the clonus self-terminating at all angles but the frequency is halved close to neutral compared with a year before. The twitch-time to joint angle curve 18 months after heel-cord lengthening has shifted upwards and is now closer to that of the nonparetic limb.

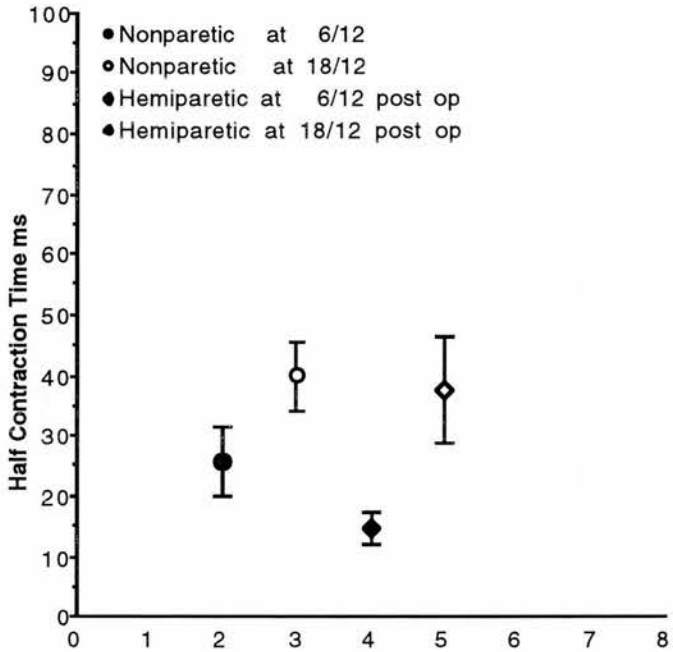
Figures 8.4.2a-f illustrate the changes over the year in **a.** reflex EMG, **b.** peak twitch force (N), **c.** 1/2 contraction time (ms), **d.** 1/2 relaxation time (ms, **e** twitch time (ms), and **f.** twitch frequency (Hz) respectively. The absolute peak force is still weaker 18 months post-operatively than that of the nonparetic limb which itself is unchanged. The most dramatic change is the return towards normal of the temporal variables of the 1/2 contraction,1/2 relaxation and twitch time times and the twitch frequency which have all slowed down significantly in the hemiparetic limb 18 months post-operatively..

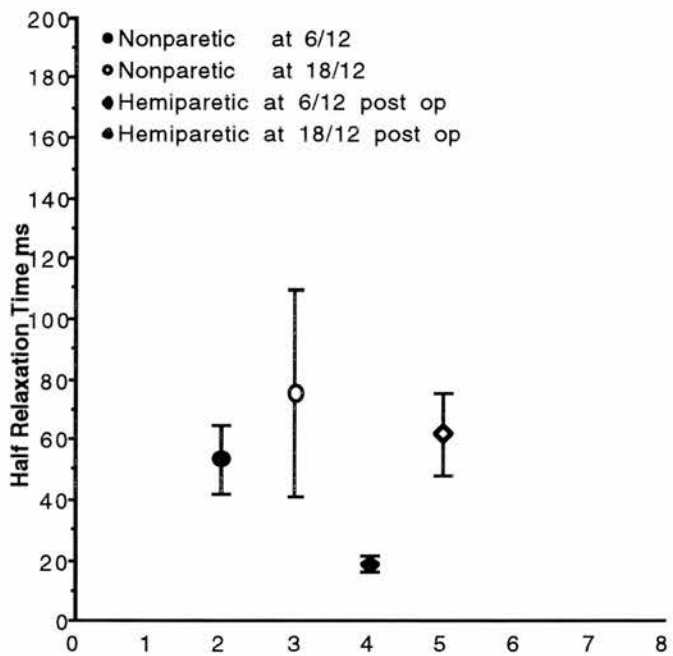
Figure 8.4.3 shows the dramatic reduction in clonus frequencies of the hemiparetic muscle 6 and 18 months post heel-cord lengthening against joint angle.



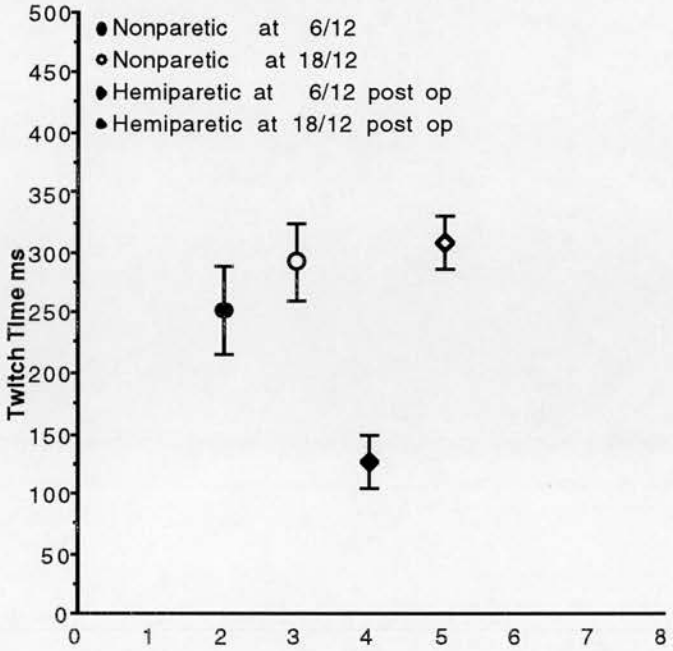


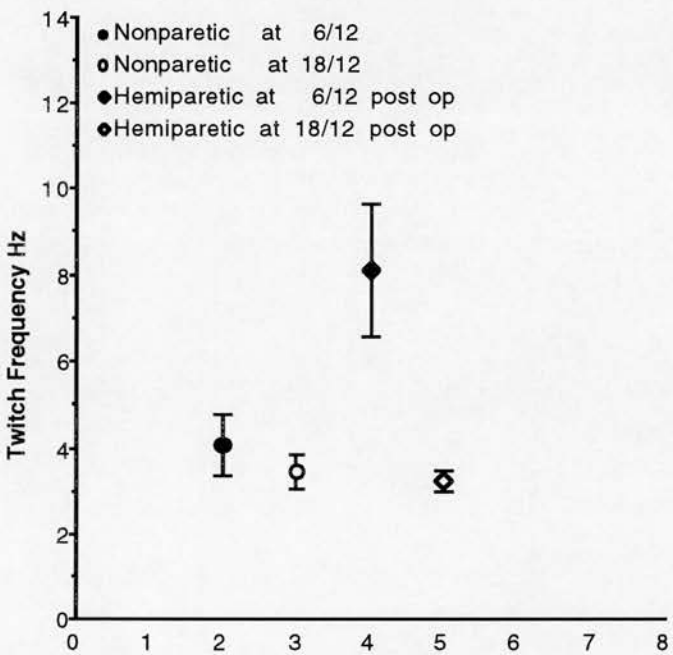
Figures 8.4.2 a-b Post heel-cord lengthening reflex twitch parameters.
Mean and standard deviations from the case in fig 8 4.1.
Top: **a.** reflex EMG (mV). bottom:**b.** reflex twitch force (N).





Figures 8.4.2 b-c Post heel-cord lengthening reflex twitch parameters.
Mean and standard deviations from the case in fig 8 4.1.
Top: **c.** 1/2 contraction time (ms), Bottom: **d.** 1/2 relaxation time (ms)





Figures 8.4.2 d-e Post heel-cord lengthening reflex twitch parameters.
Mean and standard deviations from the case in fig 8 4.1.
Top: **e.**reflex twitch time (ms) and Bottom: **f.** twitch frequency (Hz) at 6 and 18 months after heelcord lengthening. Note the slowing in all the temporal characteristics of the hemiparetic muscle 18 months after surgery. The muscle continues to be weak in absolute terms when compared to the nonparetic limb whose force and temporal characteristics change little over the same period. For reflex EMG, twitch force, 1/2 contraction and 1/2 relaxation times see figure 8.4.2.a-d, above.

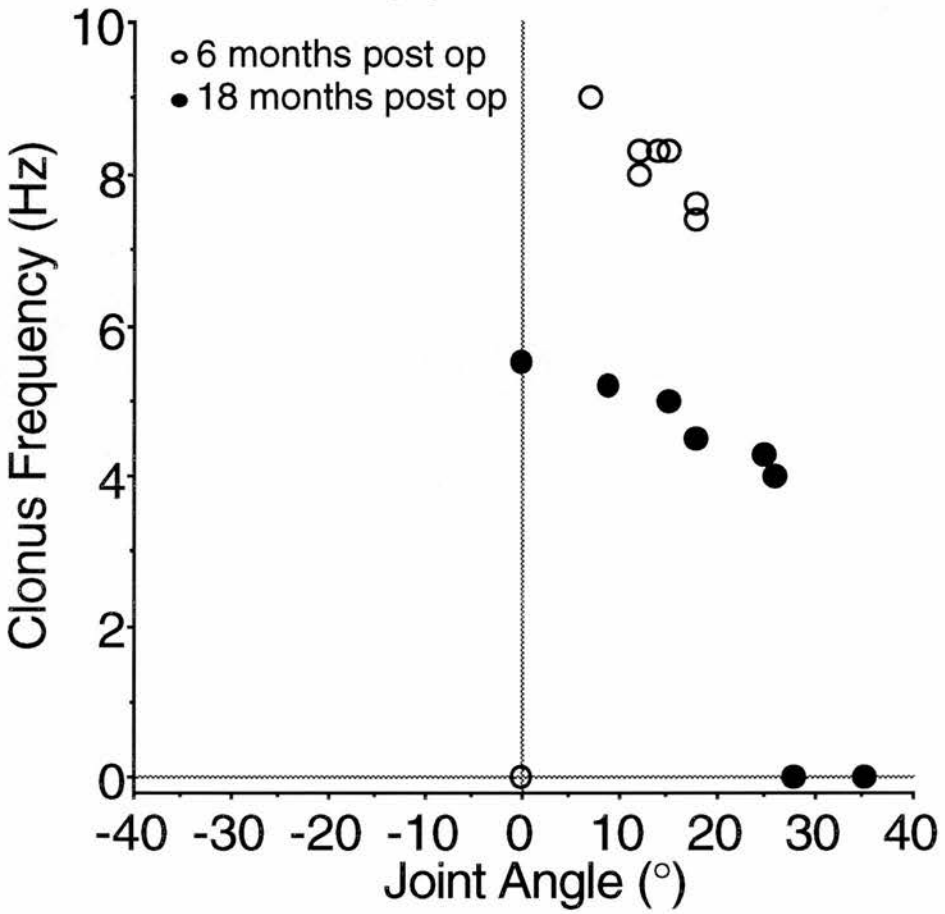


Figure 8.4.3 Clonus frequency and joint angle at 6 and 18 months after heelcord surgery.
Changes in absolute clonus frequency against joint angle six months and eighteen months after heelcord lengthening of the hemiparetic limb.
Clonus frequency can be slowed down by passive stretching in the short term 6 months and 18 months post-operatively, and is almost halved in the long term due, apparently, to changes in muscle phenotype.

8.5. Soleus reflex twitch characteristics after serial plaster immobilisation at neutral

Three children had measurements before and after plaster immobilisation at neutral for an equinus gait.

The first example comes from a five year old boy with a congenital left hemiparesis whose hemiparetic calf was studied before and after two weeks of plaster immobilisation at neutral. Figure 8.5.1a shows the changes in twitch time before and after immobilisation. It is important to state that passive dorsiflexion was only possible to -10° of plantarflexion prior to casting whereas this increased to +10° beyond neutral after casting. There is a striking fall in twitch time after casting from 400-550ms to 300-350ms across the newly acquired joint range of -20° to +10°: immobilisation has transformed the muscle twitch characteristics from slow to fast twitch, indicating that after immobilisation the joint angle-twitch time curve appears to be shifting downwards and into dorsiflexion.

In the second case, plaster immobilisation was used for four weeks in a five year old girl with a congenital right hemiparesis secondary to intraventricular haemorrhage of prematurity associated with shunted hydrocephalus, measurements being taken before and after plastering. Figure 8.5.1b shows that the twitch time before plastering (closed circles) increases steeply from 200 ms at the resting angle up to 475ms with dorsiflexion beyond neutral, whereas after immobilisation the twitch time (open squares) only varies from 200 to 275 ms for a similar dorsiflexion beyond neutral.

It is noteworthy that the nonparetic twitch times (closed triangles) do not differ from the precasting hemiparetic values. The joint angle-twitch time curve is less steep after immobilisation and appears to be shifting downwards and into dorsiflexion. Prior to immobilisation, the hemiparetic limb did not exhibit evidence of clinical clonus (fig 8.5.2a), but the twitch time increased with 15° of dorsiflexion beyond neutral (fig 8.5.2b). When measurements were repeated immediately after the cast was removed four weeks later, 5 beats of clinical clonus are evident after the initial tendon tap reflex twitch at the neutral angle as shown in figure 10c. Note, that in comparison with the brief, near synchronous, large amplitude, post tendon tap reflex EMG discharge, clonus beats are generated by low amplitude bursts of EMG discharges, each lasting approximately 25-30ms. As well as ceasing after five beats at the neutral angle, the clonus is limited to 2 very weak and slow "beats" (seen as low bumps on the force trace) at 17° of dorsiflexion (fig d). When measured at

comparable angles of dorsiflexion beyond neutral, the reflex twitch force is greater but the twitch profile appears faster after four weeks of immobilisation. This appears to favour the elicitation of clonus at the neutral joint angle in the first instance, which in turn can be inhibited by passive dorsiflexion (muscle stretch).

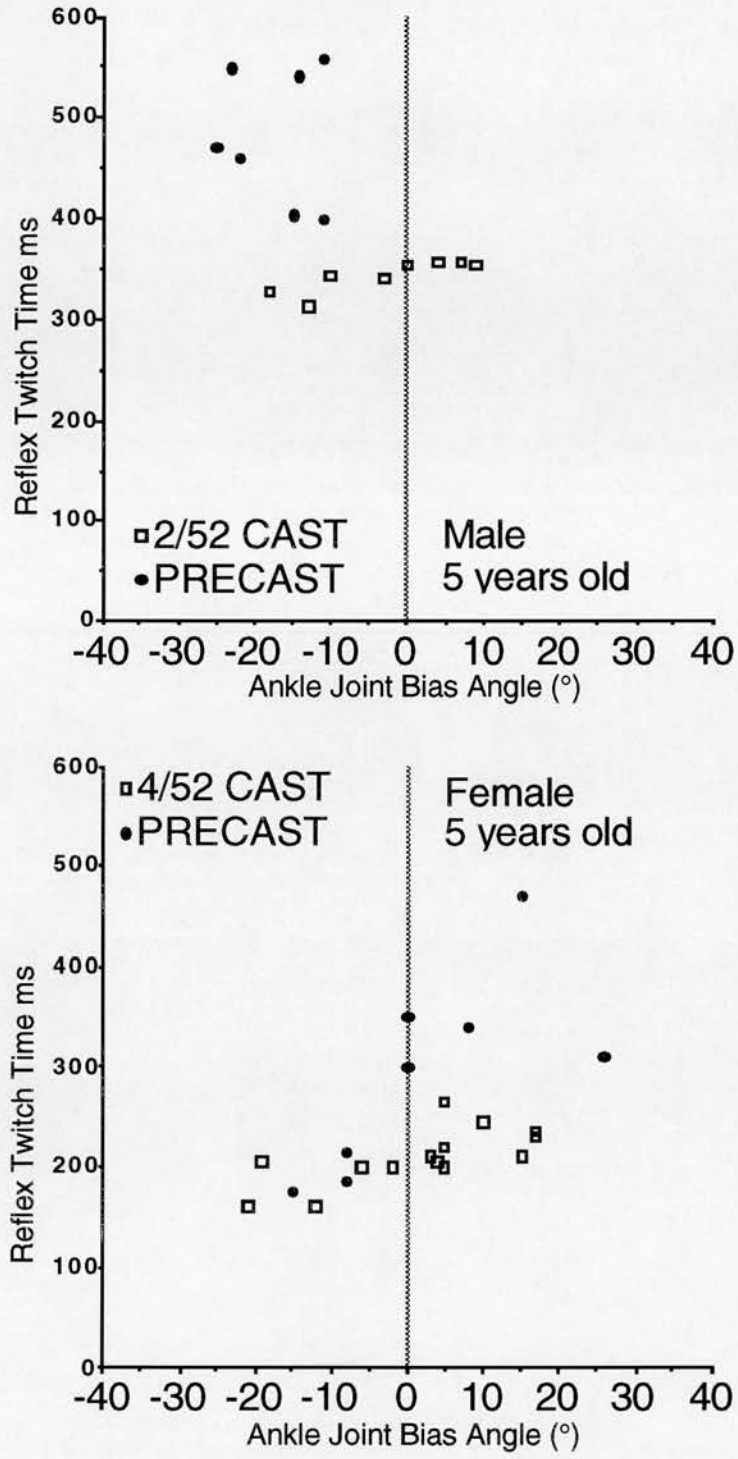


Figure 8.5.1a-b. Serial plastering to relieve equinus and reflex twitch time.
Effects of plaster immobilisation (casting) on twitch time in:
a. Five year old boy with a left hemiparesis before and after two weeks in a plaster cast.
b. before and after four weeks of casting in a five year old girl with a right hemiparesis. In both cases the reflex twitch time is faster post immobilisation and the muscles can now be dorsiflexed beyond neutral. The joint angle twitch time curve is shifted downwards and to the right. The nonparetic muscles of either case did not change their temporal characteristics over the same period.

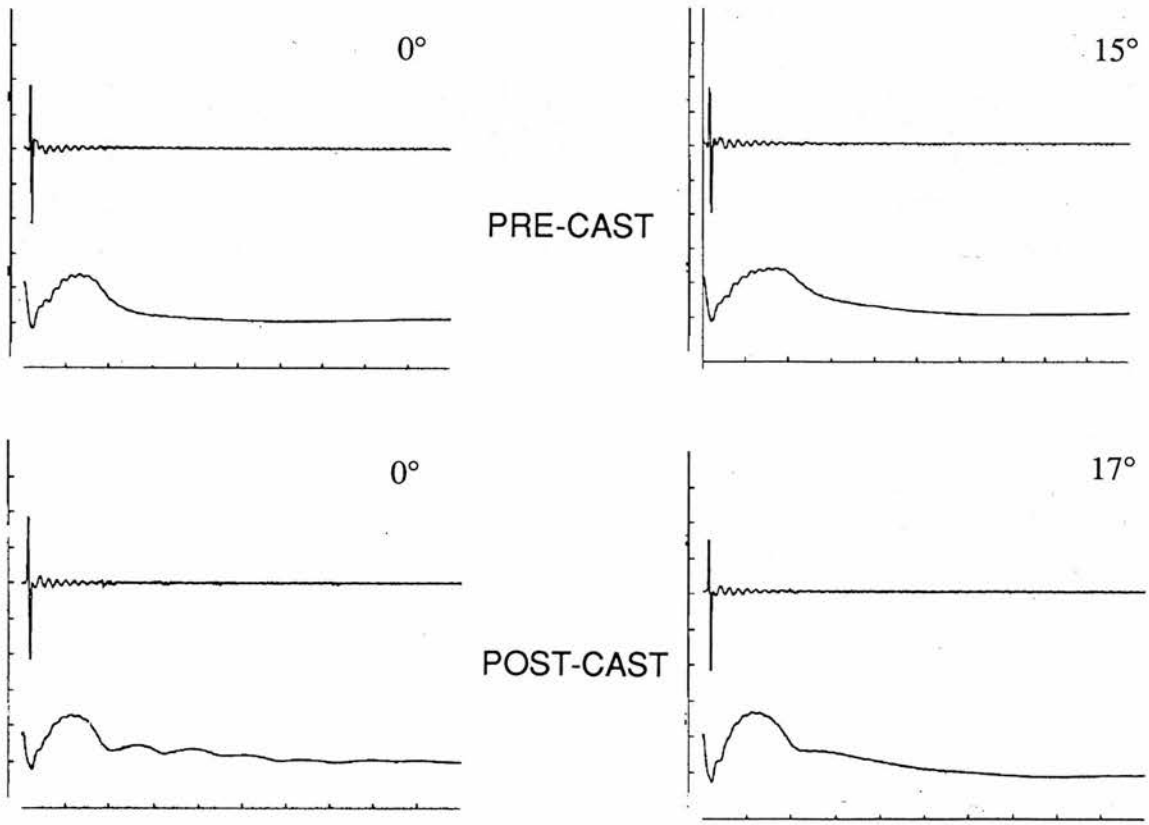


Figure 8.5.2a-d. Clonus after one month of plaster immobilisation to relieve equinus.
Physiological traces before and after immobilisation for the second case, a five year old girl with a right hemiparesis (see figure 8.5.1b).
Top: pre-cast, at **a.**0° and **b.**15°. **Bottom**: postcast, at **c.** 0° and **d.** 17°. There is evident clonus after casting (bottom left), the frequency of which can be slowed by passive dorsiflexion (bottom right).

8.5.3 Serial follow-up after serial casting to relieve an equinus gait.

The third case is a 9 year old boy with a congenital right hemiparesis and persistent toe strike in gait in whom measurements were made before plaster immobilisation and at each plaster change at 2, 4 and 6 weeks respectively.

The effects on the soleus twitch time against incremental joint angles at each fortnightly evaluation are given in figure 8.5.3a for the nonparetic and 8.5.3b for the hemiparetic limbs respectively.

Prior to casting, the hemiparetic resting angle approximates -30° of plantarflexion and only maximal dorsiflexing torques produce +10° of dorsiflexion beyond neutral which produces a passive angular range of +40° (fig 8.5.3b). This compares with a resting angle of -18° of plantarflexion for the nonparetic limb which can be passively dorsiflexed to +20° or +25° beyond neutral to produce a similar passive angular range of 40°-45° (fig 8.5.3a). It can be seen that the resting and bias joint angle do not vary much for the nonparetic limb over the six week period of assessment at fortnightly intervals (fig 8.5.3a) in contrast with the hemiparetic limb, for which the range of passive dorsiflexion beyond neutral increases at each fortnightly assessment, culminating in a resting angle of -10° and a maximum bias angle of +25° of dorsiflexion beyond neutral (fig 8.5.3b).

To this extent, the aims of casting have been fulfilled, but the other striking change relates to the twitch characteristics of the hemiparetic muscle which are slowed by about 50-60ms at neutral after two weeks of immobilisation. However, these twitch times become shorter at each subsequent fortnightly assessment. Figure 8.5.3b clearly demonstrates that the soleus muscle undergoes a succession of transformations to produce a family of joint angle-twitch time curves, the axis of which, shifts downwards into dorsiflexion with each subsequent fortnight of immobilisation. The marked effect of plaster immobilisation on the twitch characteristics in this case is evident when the bias joint angle is held at neutral (0°). Whereas the nonparetic muscle twitches vary little over the six week period (figs. 8.5.3a, c), the hemiparetic muscle twitches slow down after two weeks and then speed up again as immobilisation at neutral is maintained for a further four weeks (figs 8.5.3b, d).

Figure 8.5.4 compares the soleus reflex twitches of the nonparetic and hemiparetic limbs at the neutral angle before plastering of the hemiparetic limb and at each plaster change at 2, 4 and 6 weeks, each trace representing the average of four tendon tap reflexes.

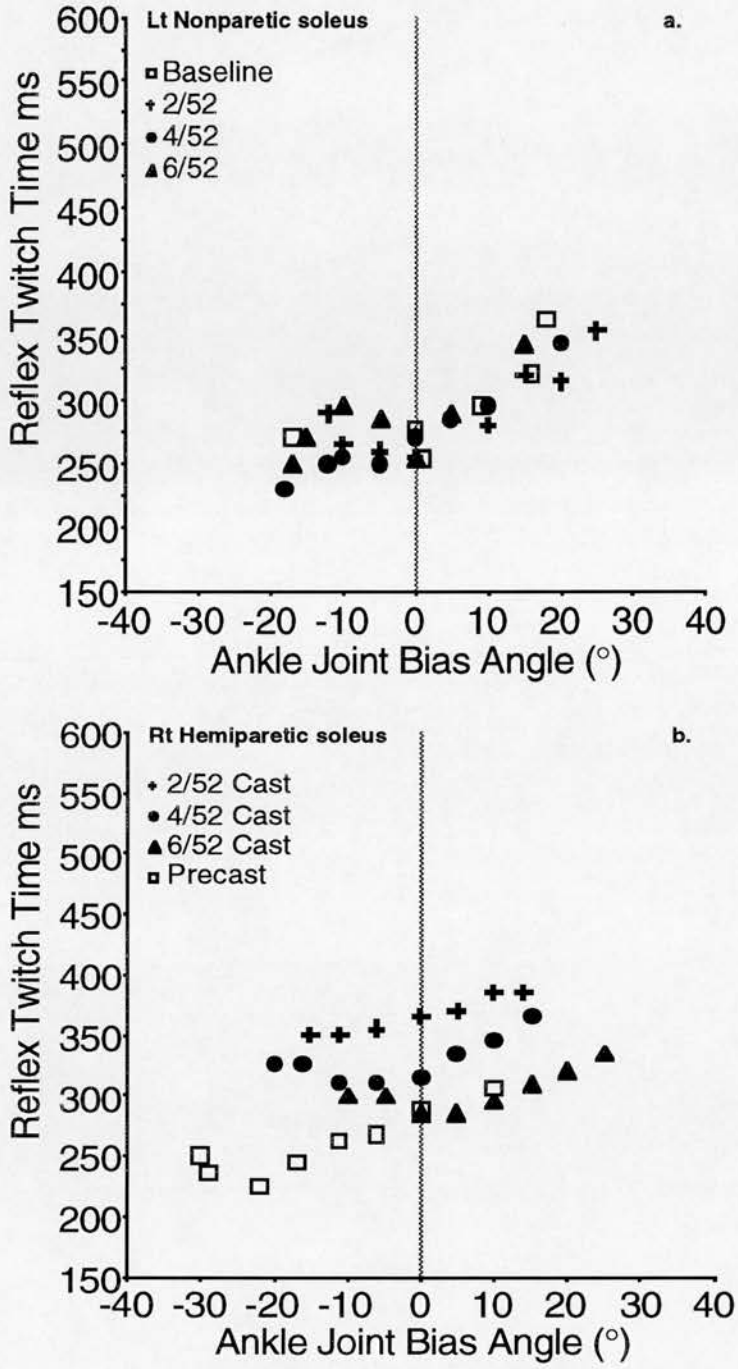


Figure 8.5.3 a-b Serial plastering over six weeks and reflex twitch time.
Effects on twitch time of six weeks of plaster immobilisation at the ankle studied before and at 2, 4 and 6 weeks after casting compared to the nonparetic limb at equivalent intervals **a**. nonparetic limb twitch time, **b**. hemiparetic twitch time, across the joint range respectively Note: **a.** that the nonparetic muscle twitch parameters do not alter over the 6 week period. **b.** The hemiparetic soleus begins in extreme plantarflexion (open squares), achieves a balanced joint range about neutral after two weeks but adopts a very slow twitch time (crosses), but as the same muscle continues to be immobilised at neutral, the joint angle shifts further into dorsiflexion (closed circles, then triangles) and the twitch times get progressively faster. Note the precast soleus twitches are fast, because the muscle is held in equinus.

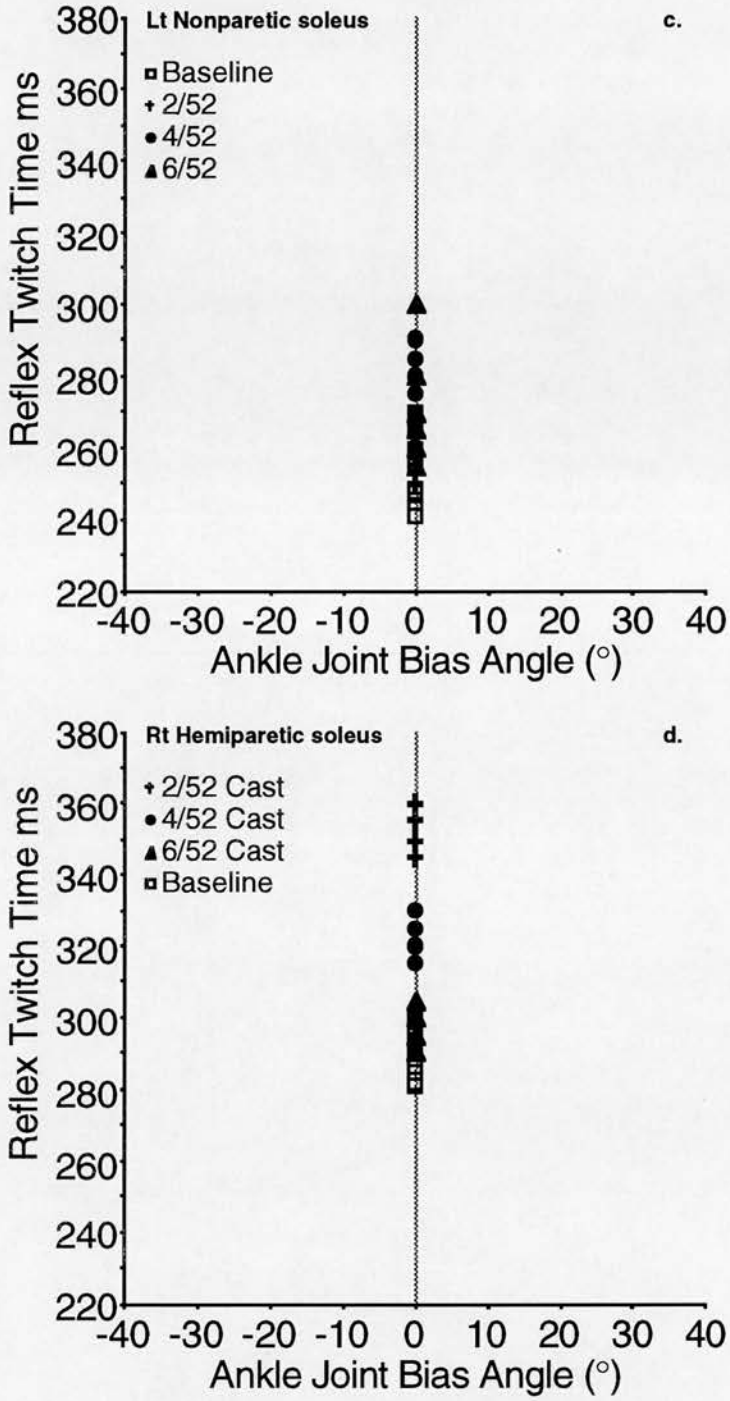


Figure 8.5.3 c-d Serial plastering over six weeks and reflex twitch time.
Effects on twitch time of six weeks of plaster immobilisation at the ankle studied before and at 2, 4 and 6 weeks after casting compared to the nonparetic limb at equivalent intervals **c**. nonparetic twitch time and **d**. hemiparetic twitch time when measured at the neutral joint angle respectively. Nine year old boy with a right hemiparesis. The twitch parameters pre- and post casting are clearly evident. Note that the nonparetic limb has a faster muscle twitch phenotype than the hemiparetic limb pre- or post-casting.
For variation in twitch time across the joint range, see 8.5.3 a-b, above.

Figure 5.5.4 Serial plastering and reflex twitch parameters.

see overleaf.

Physiological traces of reflex EMG and twitch force at neutral before and after serial casting immobilisation at neutral for six weeks measured at fortnightly intervals (case as in fig 8.5.3). Each trace represents four consecutive tap averages with time for the system to reset between taps.
Nine year old boy with a right hemiparesis.
Vertical scale: EMG = 1mV, Force = 6.37N. Horizontal scale = 100ms.

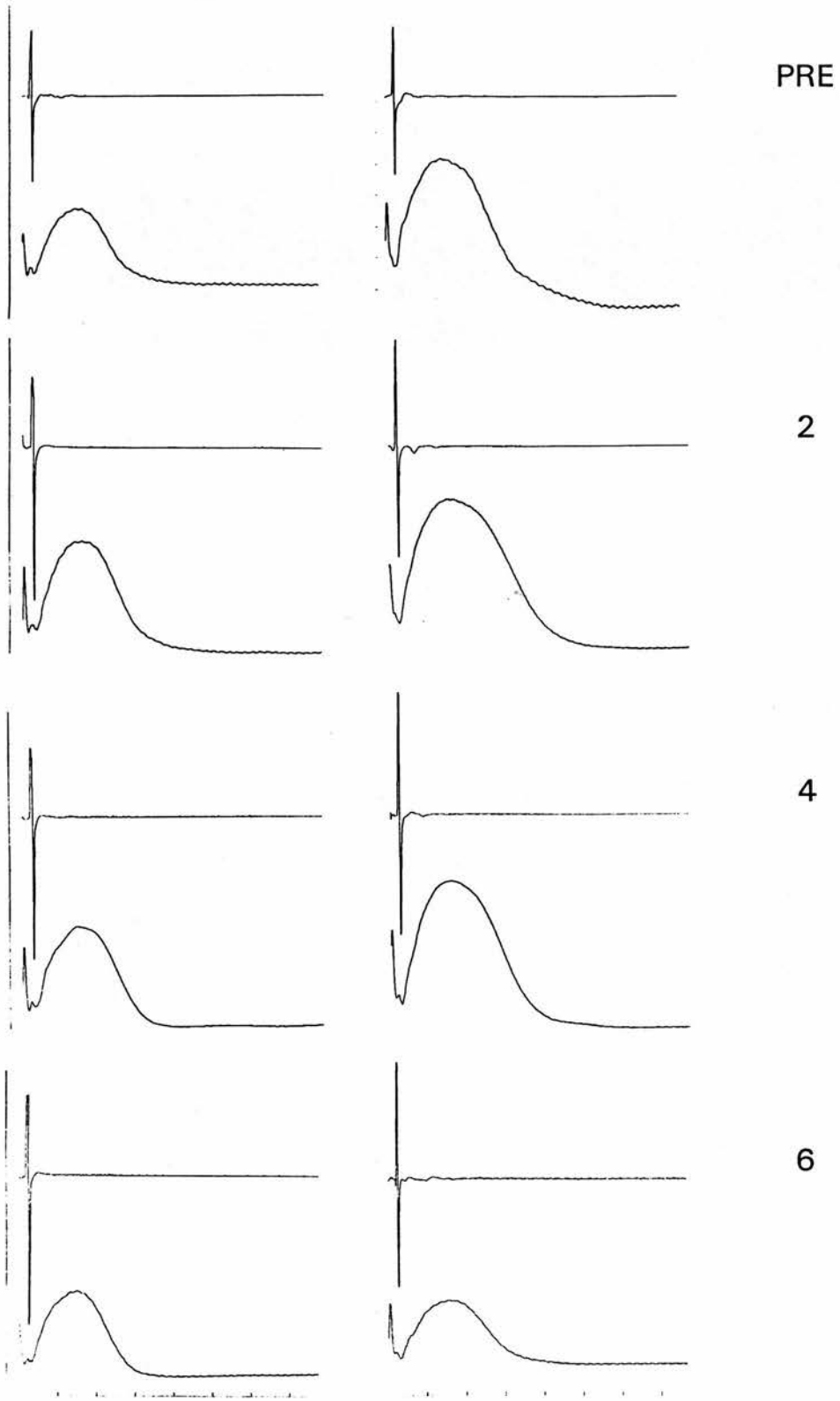


Figure 8.5.4 Serial plastering and reflex twitch parameters.

Whereas the twitch characteristics of the nonparetic limb vary little over this period, the twitch of the hemiparetic soleus strengthens and slows after a fortnight of plastering, becoming weaker and faster after each further fortnight of plaster immobilisation at neutral.

Not only is the hemiparetic soleus muscle weaker after six weeks of immobilisation than at the outset, but it has also become weaker than the nonparetic muscle, whereas prior to plastering, the reflex peak force of the hemiparetic limb was stronger than that of the nonparetic soleus muscle. The family of joint-angle to twitch-time curves in figure 8.5.3a for this case indicates that further immobilisation for a total of eight or ten weeks would have a profound effect on calf muscle physiology and function. From figures 8.5.3 and 8.5.4, the cumulative effects over time of serial plaster immobilisation at a given joint angle illustrate the direct effects on muscle physiology which had been hinted at from the two previous follow-up studies on the effects of immobilisation for only two weeks (fig 8.5.1a) or only four weeks (figs 8.5.1 and 8.5.2).

The muscle twitch phenotype appears to depend on the angle at which the muscle is immobilised and the resting tension within the muscle during the period of immobilisation. The initial marked slowing of the twitch characteristics after the first fortnight of plaster immobilisation in figure 8.5.3b and 8.5.3d is undoubtedly due to the muscle being under tension during this period (see discussion) which effectively imposes work on the muscle which in turn changes to a more forceful and slow twitch phenotype. At the same time, the imposed stretch results in sarcomere addition,since the resting angle and total passive joint range shift to the right. With prolonged immobilisation at neutral, further plastering merely immobilises the muscle at neutral *without imposing tension* on the muscle and this is the same as imposing disuse which brings about a fast twitch phenotype and eventually muscle weakness.

If immobilisation is not accompanied by any significant amount of tension, then the muscle phenotype will become faster through disuse (figs. 8.5.2, and 8.5.3). These studies raise the question of how to control for the initial tension imposed on the muscle by immobilisation at neutral, since this will vary considerably from case to case and limb to limb. An obvious corollary to this problem being how can the strength of the calf muscles be preserved while at the same time restoring a functional joint range at the ankle?

8.6 Discussion: reflex excitability in health and diseases, with and without treatment..

8.6.1 A continuum of reflex excitability.

The present study examines the differences between the reflex neuromuscular output of healthy, nonparetic and hemiparetic limbs in children of all ages up to mid adolescence. Although the experimental conditions are highly artificial, the insight into motor physiology and pathophysiology appears to provide a unifying framework which encompasses normal voluntary and reflex neuromuscular control, the natural history of reflex excitability in the hemiparetic limb and factors which may alter that natural history. Central to the conclusions of these studies are the insights into the mechanisms of ankle clonus, which can be viewed not only as an incidental by-product of cerebral (or, in other cases spinal) damage, but also as a marker of neuromuscular change over time.What happens to the limb over the course of a lifetime appears to influence the expression of neurological function.

The effects of short-term changes in muscle length on soleus muscle reflex twitches have been demonstrated in healthy and hemiparetic limbs. Passive stretching from the resting angle to just beyond neutral universally increases the reflex EMG discharge, the twitch force and the twitch time. Further stretching to maximum dorsiflexion profoundly inhibits the reflex EMG discharge and twitch force whereas the twitch time is prolonged still further.

This data resolves an apparent contradiction in the literature in which it appeared that muscle stretch was excitatory on the one hand (Tardieu, 1982) or inhibitory on the other (Burke and Lance, 1973): the data presented here indicates that both phenomena are true, provided the whole joint range is considered. Limbs were not tested in conditions of plantarflexion beyond the resting angle ie in positions of extreme equinus, because for control children, no reflex twitches were obtained, but as the hemiparetic data show, the joint angle-reflex twitch time curve exhibits considerable variability in behaviour and is not fixed for all time at a given level of excitability.

The evidence of these studies reveals a heterogeneous reflex twitch response within the hemiparetic population of limbs reflecting the heterogeneity of the hemiplegic population that has been alluded to in previous studies (Lin *et al* ,1992; 1994 a, b). As a result, the raw and normalised data for the hemiparetic and nonparetic limbs is not as tightly grouped across the joint range as that of control children or adults. This was clearly evident

from the differences in twitch characteristics between clonic hemiparetic limbs which exhibited fast muscles of varying strength in relative dorsiflexion as opposed to non-clonic hemiparetic limbs which were strong, slow and relatively plantarflexed. Neither are these differences absolute, since external interventions such as casting or heelcord lengthening and the physical biography of the limb, can alter the joint angle-reflex excitability curves.

The significance of these findings is that they indicate how the central nervous system interacts with the peripheral nervous system, and how each influences the clinical expression of the other to produce a *continuum of reflex excitability* responses which can change within a subject over the short-term eg between plantarflexed and dorsiflexed joint angles beyond neutral, or in the long term, depending on the intervening influences on the limb. The pathological response can now be interpreted as part of this continuum and the likely behaviour of the hemiparetic limb after a given intervention, or in fact any limb, becomes more predictable if the neuromuscular physiology is considered.

Herman (1970) studied 200 adults with hemiplegia which he divided into four groups according the ankle tendon jerk response, the presence or absence of resistance to passive stretch, the extensibility of the muscle and the presence of clonus. Group I hemiplegic patients, who were in the "atonic phase", had no elicitable tendon jerks, minimal resistance to stretch, completely extensible triceps muscles and no clonus. Group II patients had mild to moderately brisk ankle jerks, normal to slightly increased resistance to stretch, complete triceps extensibility and clonus could at times be elicited. Group III patients had tendon jerk hyperreflexia which was maximal at "physiological extension" (0° to -10° beyond neutral) and in addition, resistance to passive stretch was marked, extensibility was either complete or only slightly reduced, and clonus was invariable. In group IV adults with hemiplegia, the tendon jerk was just elicitable or absent at the neutral joint angle but most marked in plantarflexion, resistance to stretch was marked, extensibility was incomplete or a fixed contracture was evident and clonus was either barely elicitable or absent altogether.

Herman noted phasic "after-discharges" during the relaxation phase of the tendon tap reflex in group II and III hemiplegics, being most pronounced in the group III cases. He was able to describe that the interval between the tendon tap EMG and the first after-discharge varied according to the twitch time of the muscle. The speed of relaxation of the group III muscles was faster than that of group II hemiplegics and in addition, after-discharges were

seen most often at -30° of plantarflexion for the gastrocnemius muscles compared with 0° for the soleus muscles. Such findings in an adult hemiplegic population support the present findings in children: Herman's group II and III subjects corresponding the "clonic" children and the group IV adults to the non-clonic hemiparetic children.

There does appear to be a continuum from the inexcitable-flaccid state, immediately after injury, through to an excitable and hyperexcitable state within the first year (Thilmann *et al*, 1991), to progressively inexcitable with peripheral muscle transformation and a tendency to slow twitch muscles (for reviews see discussion below and Dietz, 1992; Forssberg and Dietz, 1997).

8.6.2 Loading and unloading muscles, muscle length and muscle speed.

The present small study does demonstrate change over time which cannot be equated with a change in the nature of the cerebral injury but is only explicable in terms of intramuscular changes which in turn affect neuromuscular coupling. The behaviour of this neuromuscular coupling can be understood according to whether the muscle is made to work or to rest.

Growth of muscles when stretches or shortening if immobilized in a shortened ste was demonstrated by Williams and Goldspink (1971 and 1973) and by Tabary and colleagues (1972). Goldspink and colleagues have studied the influence of immobilisation of muscle in the shortened and lengthened states on muscle sarcomere addition and protein turnover and demonstrated that the a 25% increase of sarcomere numbers occurs when immobilised in the lengthened state compared to a 30% decrease in the number of sarcomeres if the muscle is immobilised in the shortened state: and this sarcomere addtion is independent of muscle innervation (Goldspink *et al*, 1974). The influence of mechanical stimuli on the 'malleability of the motor system' has been reviewed:

"As a result of exercise and growth, force production is increased by an increase in the cross-sectional area of the fibres. This is associated with changes in the rate of synthesis and degradation of muscle proteins which lead to build up of the myofibrils. These then split longitudinally when they reach a critical size. This process is repeated so that the number of myofibrils increases very considerably. Also, during growth, the displacement is increased by increasing the length of the muscles. To do this more sarcomeres are produced in series along the along the length of the fibres. This is induced by stretch which also encourages fibre growth in girth as well as in length. Yet another way of changing the power output of a muscle is to change the types of muscle fibres (motor units) within the muscle. Fibre-type transformation has been shown to occur with cross innervation and stimulation but it does not usually occur with exercise training. It has been possible , however to change the fibre type proportions in young animals. Also, by combining stretch with stimulation, it has been possible for instance to make the fast glycolytic fibres add on glycolytic fibres add on fast

oxidative type sarcomeres or even slow oxidation type sarcomeres."

Goldspink, 1985, p 375.

The inherent malleability of muscle is shared by other structures:

"Perhaps the most adaptable cell are those of tissues which produce or are subjected to mechanical stress. For example, the cortical thickening, trabecular density and curvature of bone tissue are known to be determined to a large extent by the intermittent dynamic loads that occur during locomotion. the degree of keratinization of skin is determined by the extent of surface friction as well as by location, so skin cell too can detect mechanical stimuli. Fibroblasts also are apparently responsive to mechanical factors as connective tissue within muscles is increased in thickness in response to overload. Of all the tissues, skeletal muscle is probably the most adaptable. Unlike the other tissues that respond to mechanical stimuli, muscle fibres create the mechanical stresses as well as responding to them, so it is sometimes difficult to discern which is the cause or the effect when studying muscle adaptation. This is particularly true for growing muscle where some of the changes may be regarded as adaptation whilst others may be changes that are strictly programmed for in the DNA of the cell (Whalen, 1985)."

Goldspink, 1985, p376.

Exercise, inactivity and starvation has different effects on different muscles:

"An increase in the cross-sectional area and total number of myofibrils occurs during growth and during certain types of exercise training. the maximum force production of a muscle is related to the myofibril cross-sectional area so that the physiological significance of this type of malleability is apparent. However, we still need to ask what biochemical changes are occurring in an overloaded muscle which cause it to respond by producing more myofibrillar proteins so that more myofibrils can be assembled. There are two main ways proteins can be accumulated during growth or exercise training. One way is to increase the rate at which proteins are synthesized. The other is to decrease the rate at which they are broken down. Even in adult muscle proteins are constantly being synthesized and broken down and the turnover, or half-life, of the contractile proteins is of the order of 7-15 days. The soluble sarcoplasmic proteins have even shorter half-lives. A process in which more than half of the contractile proteins are broken down and replaced every 7 days or so would seem to be rather wasteful. however, it does enable the muscle to replace damaged proteins and confers on the muscle a certain adaptability for changing the the type of protein at certain stages of development and under certain physiological conditions."

"Changes in the balance between protein synthesis and degradation resulting from exercise training were investigated using a method in which young rats had to jump for their food (Watt, Kelly, Goldspink and Goldspink, 1982). This caused considerable hypertrophy of the hindlimb muscles (approximately 30% increase in muscle fibre cross-sectional area in the soleus and extensor longus muscles). The animals were exercised for 1 month before measuring the changes in synthesis and degradation rates. In extensor digitorum longus muscle, which is made up of predominantly fast fibres, the accumulation of muscle proteins occurred mainly as a result of an increased synthesis rate although the degradation rate was slightly decreased. Interestingly, the soleus muscle, which is a slow contracting postural muscle, exhibited a different strategy. In this case it was the degradation rate which significantly changed whilst the synthesis rate was only slightly elevated. It is known that the energy metabolism of these two types of muscle differs. It is also interesting to note that these different types of muscle respond somewhat differently to inactivity and starvation. The slow fibres tend to atrophy considerably in response to zero gravity, as occurs in space flight, and yet they are very resistant to the effects of starvation and to certain diseases such as muscular dystrophy. These differences may therefore be due to differences in their protein metabolism."

All the types of muscle fibres are capable of undergoing hypertrophy but they do not usually hypertrophy to the same extent. the fast contracting fibres are recruited only

infrequently (for rapid power movements or high intensity isometric contractions). When they are recruited and 'overloaded', they tend to undergo hypertrophy very readily. Therefore, the cross-sectional area of the fast fibres is increased even though the total number stays the same. This means that there are more fast-type sarcomeres in series and in parallel and hence the muscle is not only capable of producing more total force but is capable of developing that force more rapidly. Selective hypertrophy of the fast fibres can thus be regarded as adaptation for increased power production under certain situations when all or most of the fibres are being recruited."

"The slow fibres may also increase in size as a response to very frequent recruitment, but to lesser extent than the fast fibres. In repetitive low-intensity exercise, the fast fibres may hardly ever be recruited. Under these conditions they may atrophy at the same time as the slow fibres are undergoing some slight hypertrophy (for example long duration treadmill running or long-distance cycling. Thus there is a selective response depending on the type of training."

"The other way muscle fibres respond to repetitive type training is to produce more mitochondria and oxidative enzymes. Accompanying this there is also an increase in the number of capillaries per fibre (see Hoppeler and Lindstedt, 1985). In our experience both the slow-contracting and the fast-contracting fibres may respond by increasing their oxidative enzyme levels (Goldspink and Waterson, 1971). This type of mutability therefore confers increased fatigue resistance on the muscle and hence an increased aerobic power."

Goldspink, 1985, p379-380.

Quite clearly, therefore, there is a huge repertoire of responses of muscle tissue in response to external and internal mechanical stimuli. If this potential for change in biochemical and physiological properties is considered in the light of the natural history of a muscle from a child with cerebral palsy, the final outcome can be chartered according to whether that muscle was held in a shortened or stretched state; according to whether it was exercised or was inactive or surgically unloaded. How the nervous sytem interacts at a spinal level with such a muscle depends also on the historical path of that muscle.

Table 8.6 gives a prediction of the likely effects of a given intervention or natural process on neuromuscular excitability, each being classified according to whether its effects load or unload the muscles. In this context, the term "loading" is used to indicate that the muscle is made to work and "unloading" that the muscle tends towards disuse and atrophy: the term "process" refers to either an endogenous condition eg paralysis or dystonia or an exogenously imposed condition such as weightlessness or exercise.

Figure 8.6.1 illustrates the likely behaviour of the joint angle-reflex twitch time curve, according to whether the muscle is loaded or unloaded. The curve being shifted upwards after a period of loading and downwards after unloading, to the left (plantarflexion) in the presence of reduced extensibility and to the right (dorsiflexion) when extensibility is normal or increased, for instance due to sarcomere addition after serial casting or tendon elongation

after tendon-lengthening. Pathological alterations of the physiological joint range will tend to shift the curve to the left, ie in relative plantarflexion (equinus) as in the non-clonic hemiparetic limbs, or to the right , in relative dorsiflexion (calcaneous) after prolonged immobilisation at neutral or heelcord lengthening as in the clonic limbs. The therapeutic goal of any intervention would be to keep the excitability curve balanced about the neutral angle without inducing fast muscle fibre transformation or overlengthening.

Table 8.6 Peripheral factors influencing the continuum of reflex excitability.
Physical processes affecting muscle twitch characteristics and the expression of reflex excitability and clonus.

Process	**Intervention / conditioning**	**Predicted Outcome**
Loading Muscles		
Exogenous		
	i. Stretching	Muscles become
	ii. Exercise	strong and slow but
	iii. Electrical stimulation	relatively inexcitable:
	iv. Immobilisation	clonus is reduced or
	(with tension)	abolished because .
Endogenous	v. Dystonic states	relaxation is slow.
	vi. Contracture	" "
Unloading Muscles		
Exogenous	i. Weightlessness	Muscles become weak,
	eg space travel	fast and hyper-
	ii. Disuse/ lack of exercise	excitable: clonus may
	iii. Immobilisation	become prominent.
	(without tension)	because relaxation is fast.
	iv. Heel-cord lengthening	" "
Endogenous	v. Central paralysis	" "

Fast-twitch muscles may cause Ia afferent depolarisation during the relaxation phase. The speed of relaxation of the muscles may be capable of reaching the threshold firing velocity to depolarise the alpha-motorneurone pool. I f there is reduced pre-synaptic inhibition, an induced fast twitch phenotype, such as after prolonged immobilisation or tenotomy may allow the relaxation velocity to reach the reflex threshold.

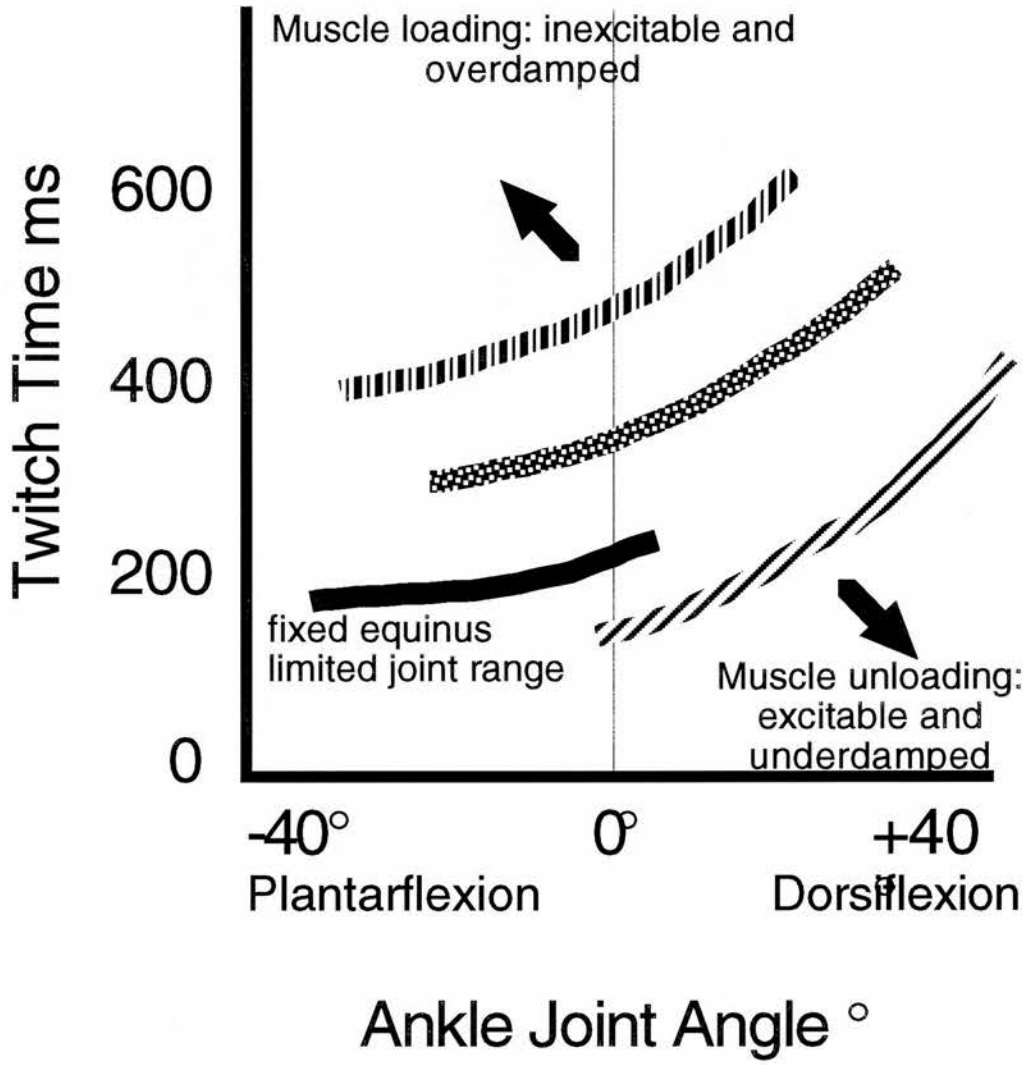


Figure 8.6.1. Natural history, treatments, soleus reflex twitch time and the joint angle
Effects of loading and unloading muscles on the functional joint range and reflex excitability. Theoretical model of muscle twitch time against joint angle curves. Reflex excitability and the emergence of clonus may be seen according to whether the twitch time is fast and the joint range is shifted towards dorsiflexion. Conversely, reflex inexcitability emerges as the twitch time slows and the joint angle shifts towards equinus.

8.6 3 Mechanisms of clonus.

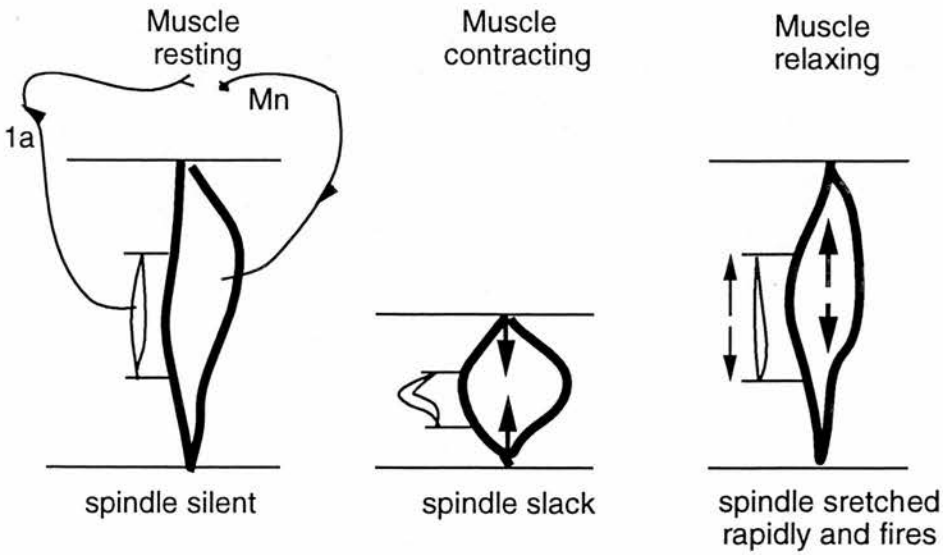
The emergence of pathological clonus can be seen as a combination of a failure of normal physiological control of muscle twitch characteristics, a loss of spinal presynaptic inhibition and an abnormal twitch time-to-joint angle profile. But even under conditions favourable to clonus, muscle stretch has a modulating effect on the muscle strength, twitch speed and most importantly the clonus frequency which was reduced with successive stages of passive dorsiflexion beyond neutral. This was seen in all four hemiparetic cases with clonus and the further case in whom clonus emerged after a month of plaster immobilisation at neutral.

The halving in clonus frequency eighteen months after heel-cord lengthening is matched by an increase in reflex twitch strength and slowing of the soleus twitch time. As presented, the component of the total twitch time which is particularly affected by passive stretching is the relaxation time, which increases markedly with dorsiflexion beyond neutral. These observations are in keeping with those of Herman (1970) and Szumski et al, (1974) who described the importance of muscle contraction and relaxation patterns in the generation of clonus. another peripheral modulator of the clonus frequency was provided by lansek (1984) who demonstrated a change in clonus frequency with reflex path length duration. Such observations provides an important clue to the pathophysiology of clonus in which the peripheral characteristics of the neuro-muscular system play a crucial part.

Figure 8.6.2 illustrates the role of muscle relaxation time in the phenomenon of clonus. The muscle spindle (intrafusal fibre) lies parallel to the skeletal muscle sensing changes in length and velocity when the muscle is passively stretched, during eccentric voluntary contraction (ie actively paying out)or during muscle relaxation. In the latter case, the faster the relaxation, the faster the stretch stimulus to the spindles, the larger the afferent input along the Ia afferents to the spinal cord and the greater the likelihood of a reflex threshold being reached causing the motorneurones in the spinal cord to fire sufficiently to evoke a reflex muscular twitch, or the first clonus beat.

The studies presented, all demonstrate the slowing in muscle relaxation with increasing soleus dorsiflexion, and the examples of hemiparetic limbs with clonus show how the frequency of clonus diminishes with passive muscle stretch, presumably because in positions of increasing dorsiflexion, muscle relaxation becomes progressively slower and

eventually is incapable of providing an adequate velocity stimulus to cause a reflex muscular twitch, or the first clonus beat. According to this model of clonus and the data presented, if the twitch characteristics of a hemiparetic muscle exhibiting clonus become slower, the frequency of clonus should diminish or may be abolished. Conversely, a speeding up of the twitch characteristics would result in a more readily excitable reflex (since the velocity of relaxation is faster) and a higher clonus frequency since the twitch times are faster.



The pathophysiology of clonus

The muscle spindle fires in response to rapid muscle relaxation
If muscles have pathologically fast relaxation times, the spindle will be stimulated and elicit a reflex muscle contraction which at the optimal joint angle (muscle length) causes a self-renewing reflex: clonus.

Figure 8.6.2 Model for muscle twitch characteristics, spindle stretch and clonus.
The closed loop model of reflex excitability. The extrafusal muscle plays a peripheral modulating role on reflex expression, notably the emergence of clonus.
The sequence may renew itself, provided the speed of muscle relaxation keeps reaching the reflex velocity threshold.

The most significant short-term determinant of clonus frequency appears to be the muscle length (or joint angle), which modulates the muscle twitch time and the reflex excitability. The evidence presented confirms that the motor twitch profile absolutely limits the clonus rate.

8.6.4 Attempts to modify the frequency of clonus.

It is generally agreed, that the pathological clonus frequency falls narrowly between 4-8Hz (Dimitrijevic *et al*, 1978) and that physiological clonus, brought about by driving sinusoidal torques has a similar frequency of 5-8Hz (Gottlieb and Agarwal, 1977; Rack *et al*, 1984).

There have been a number of unsuccessful attempts to alter the frequency of clonus in a variety of limbs by increasing the inertia of the limb, which only reduces the amplitude of the clonus beats (Walsh, 1971) . Walsh (1976) was also able to show in a case of traumatic paraplegia, that the clonus EMG discharges may persist even in the absence of an observable mechanical movement of the limb involved although fluctuations in the skin overlying the calf muscles could be seen at this time which must reflect weak isometric muscular twitches not capable of moving the foot plate.

This evidence was taken to support the notion of a central clonus oscillator or pacemaker (see also Dimitrijevic, Nathan and Sherwood, 1980). More recently, topical xylocaine applied to the leg and calf was shown to reduce the amplitude, but not the frequency of pathological and physiological clonus as well as that of an exercise -induced physiological action tremor within 60 minutes of application (Mills and Pozos, 1985).

In the present study, clonus frequencies of up to 9Hz were note 6 months post-tenotomy. Under these conditions, the recordings being virtually isometric, the movement is intramuscular and restricted to the sliding in and out of the muscle fibres ie taking up "slack", which would be sufficient to unload the muscle spindles during the muscle contraction and reload them at speed during relaxation. The faster the relaxation, the greater the velocity stimulus to the muscle spindles and the more synchronous and powerful the ensuing reflex EMG discharge of the next clonus beat. Under such conditions, a muscle which is slow to relax (and stiff), offers only a weak velocity stimulus to the muscle spindles which in turn provide a more dispersed, weakly depolarising signal to the spinal motor neurones of the anterior horn.

Successive clonus beats become weaker as the intramuscular relaxation becomes progressively slower and eventually falls below the reflex threshold velocity. The critical importance of the joint angle for eliciting and maintaining clonus can now be understood in the knowledge that the soleus reflex EMG of the ankle jerk is largest close to neutral, the position of maximum reflex excitability, and diminishes steeply thereafter (see also Herman 1970). Apart from the initial rapid stretch, the "trick" for maintaining clonus is to elicit clonus near or at the position of maximum reflex excitability and to maintain that position.

Clinical observations of clonus at the bedside demonstrate this phenomenon well, provided that after the initial brisk dorsiflexing jerk, the examiner maintains a constant joint angle.

Previous studies (Lin, Brown and Brotherstone 1994 a, b), which looked at surface EMG discharges and the velocity of stretch following abrupt thigh and calf muscle stretches at different joint angles, clearly demonstrated the phenomenon of the reflex velocity threshold and gain, and the inhibitory effect of muscle stretch, but failed to provide an adequate "stimulus-response" mechanism for ankle clonus (see section 6.3.8).

This presumably arose from the use of an "open loop" model of the nervous system in which measurements included the EMG discharge but not the mechanical contraction and relaxation of the muscle which followed as in the present "closed loop" model. The present study, although entirely non-invasive and painless, appears to capture the main events regulating reflex neuromuscular modulation as well as providing an explanation for the phenomenon and modulation of clonus.

In a study of clonus in subjects with spinal pathologies ranging from multiple sclerosis, hereditary spastic paraplegia (Strumpell-Lorrain syndrome) spinal transection and spinal compression, Rossi, Mazzocchio and Scarpini (1990) were able to demonstrate progressive slowing of the soleus clonus frequency leading to complete abolition of clonus some 7 minutes after partial nerve block was performed by applying a sphygmomanometer cuff inflated to 300mmHg to the upper third of the leg. The soleus tendon and Hoffman reflexes could still be elicited 27 and 28 minutes after the disappearance of the clonus following ischaemic calf compression. The authors conclude that the clonus frequency is altered in part by a direct effect of ischaemia on the muscle relaxation phase, such a direct muscle effect occurring before the ischaemia has blocked transmission of either the tendon

tap or the H-reflex along the 1a afferent fibres from the soleus muscle spindles. These same authors also clearly demonstrate that the duration of clonus varies considerably over a 30° joint range with a maximum duration of 8s at neutral falling to 0.25 s at 30° of dorsiflexion beyond neutral.

The most significant finding for therapy and rehabilitation arising from the present work and that of Rossi *et al* (1990) is the evident link between joint angle, muscle twitch speed and clonus frequency.

That clonus is more prominent in spinal rather than cerebral pathologies (Whitlock 1990) can be explained in part by the observation that spinal injury results in a greater degree of paresis and disuse of the muscles favouring a slow to fast twitch muscle transformation (Lieber, 1986b). The lag of many days or weeks in the onset of clonus following injury to the CNS can also be partly explained in terms of the time taken for the plegic or paretic muscle to develop fast twitch characteristics as a result of the immobility and disuse (unloading) brought about by the injury.

8.6.5 Reflex excitability and muscular transformation.

Reflex excitability and spasticity are known to reach a peak between two and three months following stroke and to diminish progressively over the following year (Herman 1970; Thilmann, Fellows and Garms, 1991). The tendon jerk reflex EMG may continue to increase throughout the first year following a stroke indicating that separate mechanisms may underlie the clinical expression of the short latency as opposed to long latency stretch reflexes (Fellows, Ross and Thilmann, 1993). The motor neurone pool may reshape itself, reducing the number of participating motorneurones by half to produce larger motor units with a predominantly slow muscle phenotype by some 20 months after the acute stroke (McComas et al 1973) and histochemical analysis of muscles of the hand (Young and Mayer 1982) and leg (Edström, 1972 and Dietz *et al*, 1986) have also confirmed slow muscle fibre transformation and wasting in established long-term cases (see rewiew in CP by Dietz &Berger, 1995).

In a previous clinical study exploring the mechanisms of hindfoot equinus in hemiplegia, clinical ankle clonus occurred in only 7/24 cases (Lin and Brown 1992) which accords well with the trend towards decreasing reflex excitability with time after injury. In the case of children with cerebral palsy, the post-injury period encompasses the whole life-span.

This gradual reduction in excitability towards a state of increased viscoelastic, and hence, non-electric, muscular resistance described by Foley (1960), Herman (1970), Dietz *et al* (1981 and 1983) and Hufschmidt and Mauritz (1985) may be temporarily altered (reversed or accelerated) with external interventions. Particularly in the striking case of the studies of clonus following heelcord lengthening, the rapid twitch profiles are clearly pathological when compared with those of the nonparetic limb and even other hemiparetic limbs: such twitches defy the conventional chronic changes described above, consigning such muscles to a life-long slow phenotype.

The conventional view in relation to reflex excitability is to suppose that it arises because of the severity or extent of distribution of the initial brain or spinal injury: but this does not explain the waxing and waning of the phenomena over the ensuing weeks and months following heel-cord lengthening, nor does it explain the mechanism by which this waxing and waning of reflex excitability finds physical expression. Recent work (Noth, 1992, Nacimiento *et al*, 1993) has failed to demonstrate support for the proposals advanced by McCouch and Austin (1958) for intraspinal sprouting of primary sensory afferent nerves after cerebral injury.

Nor is there evidence for an increased sensitivity to velocity of stretch of the intrafusal fibres (Burke 1983). A loss of presynaptic inhibition of impulses reaching the spinal cord from the muscle spindles (Gillies et al 1969, Burke and Ashby 1972) does explain how normal physiological speeds of muscle stretch result in abnormal reflex responses by virtue of a lowering of the reflex velocity threshold and an increase in the reflex velocity gain but the variability of clinical expression between and within subjects over time can now in addition be explained in pat, in terms of peripheral changes occurring in the muscle, the twitch characteristics of which can be transformed by a very large number of endogenous and exogenous stimuli (Buller, Eccles and Eccles, 1960; Salmons and Sreter, 1976; Pette, 1980; Vrbová ,1980; Pette and Vrbová, 1985; Lieber, 1986 a, b and c).

Vrbová (1980) has described the time course for changes in muscle phenotype to be in the order of three to six weeks, beginning with an alteration in muscle vascularisation within the first few days and ending with a transformation of the myosin ATPase properties of the muscle fibres. Changes in the sarcoplasmic reticulum calcium uptake mechanism are thought to mediate the rapid relaxation phase of tenotomised and immobilised muscles over

a two to four week period (Barry *et al* ,1994).

The time courses given for such changes in the physiological behaviour of muscles corresponds to the four to twelve week interval required to reach peak reflex excitability quoted in stroke follow-up studies (Herman, 1970; Thilmann, Fellows and Garms, 1991), and it is reasonable to suppose that the effects of acute paresis and immobility have an initial unloading effect on muscles which may pass through an initial fast-twitch phenotype, which in turn supports the expression of pathologically brisk reflexes and clonus.

These potential changes in the active properties of muscle are followed by chronic changes in the passive properties of muscle that have been described in children (Foley 1960, Dietz, Quintern and Berger, 1981) and adults (Herman 1970, Hufschmidt and Mauritz, 1985), producing muscles that offer a high biomechanical (rheological) resistance to stretch so that a year after brain injury, muscles are likely to undergo transformation to a slow fibre phenotype because they are under chronic tension (loading). Once again, muscle physiology appears to be a rate-limiting feature in neuromuscular control.

A number of detailed studies have demonstrated fast to slow muscle transformation in response to chronic electrical stimulation of immobilised muscle (Cotter et al 1986), recovery from immobilisation-induced atrophy of rabbit soleus muscles by chronic low-frequency electrical stimulation (Cotter *et a*l, 1988 and 1991). The effects of tenotomy and muscle immobilisation either separately or in combination are shown to produce dramatic biochemical and histological changes of atrophy and fast-twitch transformation in rabbit soleus muscle: immobilisation after tenotomy being capable of inducing muscle necrosis and macrophage invasion (Barry and colleagues, 1994).

This doubly deleterious effect of tenotomy and immobilisation can be entirely reversed by chronic electrical stimulation in the convalescent period or even prevented, provided the electrical stimulation is delivered to the muscle at the outset. An additional benefit of low frequency electrical stimulation in the convalescent phase is to mimic exercise which increases the cardiac output necessary to supply blood to the contracting muscles, which become richly vascularised and less fatigable (Vrbová 1980; Pentland 1993) in contrast to phasic, glycolytic muscles, which can only support brief periods of anaerobic activity and rapidly fatigue and lack endurance.

The effects of total tenotomy of all the muscles acting around the ankle joint on reflex

modulation were investigated by Vrbová (1963) in the rabbit. These studies indicated a loss of reflex activation of the soleus muscle in the conscious animal in the immediate aftermath of the surgery, while tibialis anterior reflexes seemed preserved in the initial two weeks but practically disappeared by six weeks post operatively. The results of the experiments were compatible with the established evidence that under normal physiological conditions, monosynaptic activation of slow, postural muscles such as soleus, is more readily elicited than that of phasic muscles such as the tibialis anterior. When the animals were anaesthetised however, reflex activity was greater than ever indicating that there is a central depression of reflex activation in conscious tenotomised animals with a normal CNS, but that if this central depression is abolished by anaesthesia or interrupted by spinal section, reflex activity is enhanced. Clearly there are implications for the practice of tenotomy in children with pre-existing cerebral lesions in whom the post tenotomy central depression will be lacking. However, anaesthetic agents such as halothane and isoflurane in children with cerebral palsy (Soriano *et al*, 1995; de Jong et al, 1968; 1967) have been shown to completely abolish the intra-operative H-reflex testing in cases under going selective dorsal rhizotomy, as does nitrous oxide (Soriano *et al*, 1995).

Clear differences exist between the experimental model and clinical practice since no surgeon aims to completely tenotomise a muscle, but merely to lengthen the tendon. Insufficient data exist on either traditional percutaneous heel-cord lengthening or the gastrocnemius aponeurotic lengthening (Vulpius and Stoffel 1913, see review by Rosenthal and Simon 1992) to determine how they compare in terms of preserving or enhancing motor function; increasing or reducing reflex excitability.

8.6.6 Orthopaedic practice and clonus.

Current orthopaedic practice varies worldwide: some surgeons applying casts to immobilise the limb following tendon-lengthening procedures, others use only light bandages and recommend intensive physiotherapy within days of surgery. One recent study (Comacho, Isunza and Coutino, 1996), attempted to abolish exacerbations of ankle clonus following heel-cord lengthening by concomitant neurectomy of the gastrocnemius muscle. The authors compared tendo Achilles lengthening alone in 6 children (11 limbs) with lengthening and neurectomy in a further six (10 limbs) and found that clonus subsided in only 27% of those ankles which had undergone lengthening compared with half of the limbs

which had also been neurectomised. Interestingly, the authors of this small study state :

"Treatment of equinus foot deformity in children with spastic cerebral palsy is often complicated by associated clonus. Results of casting, bracing, and surgery may be compromised by this aberrant muscle activity."

Camacho *et al* 1996

Camacho and colleagues (1996) are clearly highlighting a gap in knowledge of the precise effects of peripheral treatments on the clinical expression of abnormal neurological states. If muscles must be lengthened, muscle power and twitch profiles could be maintained after initial wound healing by concomitant exercises or chronic electrical stimulation to prevent adverse wasting and weakness (Barry *et al*, 1994).

Traditional physiotherapy concentrates on "tone reduction" by slow passive stretching which certainly has a bearing on reflex excitability if the joint angle is altered by the application of an orthosis, but less attention is paid to strengthening and exercising muscles which may be functionally more useful as well as serving to dampen reflex excitability. These studies and those of others, indicate that plaster immobilisation can produce marked changes in neuromuscular excitability so that casting is not necessarily a "benign" procedure.

All treatments must be seen as a form of exogenous manipulation of the motor system. Table 8.6 provides a predictive framework of the likely effect on reflex excitability of a variety of exogenous interventions and endogenous processes depending on whether the outcome results in *loading* or *unloading* of the muscles. Other specific circumstances may be fitted into this framework but essentially, this model of reflex excitability indicates that even conditions of prolonged weightlessness, providing the unloading is sufficient, could result in the expression of clinical clonus.

This study comprised a very heterogeneous group, typical of the contemporary hemiplegic childhood population, the clinical expression of which has been shaped by a variety of influences. It has sought to explore reflex twitch characteristics in an operationally-defined way and to demonstrate some aspects of neuromuscular integration which offers a predictive instrument for the likely effects on neuromuscular performance of certain interventions.

A better understanding of the natural history of the motor disorder of cerebral palsy and the effects of treatments will result in better tailoring of specific treatments. By stressing a continuum of reflex excitability, variations between and within subjects are open to

modification. The concept of a continuum of reflex excitability has been used to explain the phenomenon of clonus in healthy adults under certain conditions of physical testing producing muscle fatigue (Gottlieb and Agarwal, 1977, Rack, Ross and Thilmann 1984).

At the most general level, all the physical therapies (both invasive and non-invasive) currently available are likely to affect the muscles in one way or another, but clearly the aim must be to preserve muscle strength, maintain the functional joint range of the limb and promote movement. The spectrum of clinical observations and treatment effects presented here could focus future therapeutic interventions on combining the benefits of different treatment modalities to modify or offset the undesirable consequences of a unimodal approach. The fact that exogenous events have such a profound effect on neuromuscular performance should be taken in the optimistic vein that strategies can be designed to improve motor function and performance.

8.7 Summary

The electromyographic (EMG) responses to tendon taps at the ankle and ensuing soleus muscular twitch forces and temporal parameters were studied at varying angles across the joint range in 22 healthy control and 18 age-matched children with congenital hemiparetic cerebral palsy.

8.7.1 Reflex EMG and twitch force and twitch time.

In all cases, passive muscle stretch caused an increase in the reflex EMG and twitch force which peaked on either side of neutral respectively diminishing with further dorsiflexion. Twitch times increased with each dorsiflexing increment, being slowest at maximum dorsiflexion and fastest at the resting angle.

8.7.2 Hemiparetic data.

Heterogeneity of the hemiparetic data is evident when compared to nonparetic and control data with clear differences in the clonic (fast twitch) as opposed to nonclonic (slow twitch) muscles.

8.7.3 Clinical expression of clonus with joint angle and after intervention.

In four cases with clinical clonus, clonus frequency was reduced by passive dorsiflexion. Plaster immobilisation for one month brought out clonus which was previously absent in one case, and in a further two cases, caused a fast-twitch phenotype to emerge. Follow-up after heelcord lengthening in one case showed that clonus frequency decreased

as the muscle twitch strengthened and slowed. Short and long term peripheral manipulations appear to regulate neuromuscular excitability according to whether muscles are loaded or unloaded.

8.8 Conclusion.

Although damage to the nervous system provides the setting for reflex excitability, the data suggests that the joint angle (muscle length) and the muscle twitch phenotype of any given limb for any given case appear to dictate the actual speed and strength of reflex muscle twitch and clonus profiles. This illustrates how peripheral manipulations of muscles and tendons may alter the expression of what have hitherto been considered as exclusively central phenomena.

9. A Study of the Physics and Neurology of Alternating Movements

"A striking characteristic of many small animals is the rapidity of their movements, and there have been many speculations, since the days of Galileo and Borelli, on the problem of how speed and strength are related to size. The inherent strength of a contracting voluntary muscle fibre is roughly constant, being of the order of of a few kilograms per square centimetre of cross-section. The speed, however, varies enormously, a thousand-fold or more, between different muscles and and different animals. In general, the smaller muscles and the muscles of smaller animals are quicker. The elementary unit, the 'atom', of muscular activity is the so-called single twitch, the rapidly reversed response to a nerve impulse: but practically all movements, and all postures are due to contractions maintained by the fusion of successive twitches. The advantages of speed are obvious, but they must be balanced against the fact that a higher intrinsic speed entails a proportionally greater expenditure of energy in maintaining a contraction for a given time. The possibility of this great variation in speed is what has given such a wide latitude of size and function in the design of animals."

AV Hill, 1950, p450.

"The speed at which a given muscle has to work is dictated by the functions it has to fulfil and the size of the body of which it is part. a humming bird, while hovering, moves its wings about 55 times per second; while flying slowly backwards about 60 times, in forward flight about 100 times per second. The ruby throated humming bird is said to be able to fly at 50-60 miles an hour; its developed horse-power, reckoned per unit of body weight at this speed, must be enormous compared with that of larger birds and animals; it weighs about 2 grams. The sparrow moves its wings some 15 times a second and the stork only 2 or 3 times. Theses frequencies are roughly in inverse proportion to the cube roots of the weights, ie to the linear size. if the sparrows muscles were as slow as the stork's it would be unable to fly. If the stork's were as fast as the humming bird's it would be exhausted very quickly. These differences do not depend on nervous control but directly on the intrinsic qualities of the muscles, they are found equally in in isolated muscles stimulated electrically though naturally they are accompanied by corresponding differences in the nervous system. The speed, in fact, of the living moor must be designed, like that of a ship, an aircraft or a machine tool, to meet the requirements of the job.

AV Hill, 1950, p452.

9.1.Background: Limb segments posture, movement and reflexes.

Any given posture or movement depends on a continuous interaction of peripheral and central mechanisms. The physical properties of the limb segments narrowly define the range and economy of action, so that motor control begins with the anatomical arrangements of the limb segments and the physical properties of the muscles as the peripheral elements of the motor system. These in turn interact with simple and complex reflexes, postures and voluntary motor activation to produce controlled posture or movement. The endless repertoire of skilled actions which develop, which may be mastered, or learned, is undoubtedly based on a constant evolutionary honing of these peripheral and central elements.

The emergence of motor control from newborn to adult parallels somatic growth of the body and hence increases in limb inertia which impose further demands on motor

physiology. As will be seen, inertia can be controlled by posture, which modifies limb length and one aspect of control is the modulation of inertia while at the same time preventing unwanted oscillations. Movement at speed imposes great challenges on the motor system in terms of the energy consumption and force output required to overcome inertia while at the same time maintaining the accuracy and smoothness of movements. The delicate alternating movements of which adults are capable are markedly different from the motor output in newborns which begin with obligate postures, complex reflexes and mass movements which can be provoked by non-specific stimuli: the classical example being the startle response. But the exact mechanisms which underpin this emergent control are poorly understood.

The study of motor development has predominantly focussed on the maturation of the central nervous system elements such as the brain, spinal cord and peripheral nerves. Certain functions such as walking undoubtedly unfold according to a developmental plan and require no conscious learning, in contrast to a dance routine or using a pencil both of which are learned culture-dependent actions and not innate features of human motor development.

The basic reciprocating mechanisms for walking are likely to be laid out at spinal level, sharing much in common with more primitive vertebrate movements (Grillner 1986).

Nevertheless, even standing and walking change considerably with age, indicating that changing cortical inputs modify either the basic spinal patterns, or the selection of a repertoire of spinal patterns. The tendency to co-contraction and flexion synergies in infancy changes to that of a phasic interplay of muscles and the production of joint asymmetries in the second year of life which are necessary for a mature gait, a feature which fails to emerge in the child with cerebral palsy (Leonard, Hirschfeld and Forssberg, 1991).

Further examples of the mismatch between muscle activation patterns and the physical demands of standing are provided by Nashner's tilt platform experiments (1983 and 1985). In these non-invasive experiments subjects attempt to remain standing on a platform which may be tilted unexpectedly. Healthy controls react to such perturbations by activating their muscles in a *distal-to-proximal* fashion serving to stabilise the muscles and joints closest to the moving surface first. By contrast, subjects with damage to the central nervous system, tended to activate muscles in a *proximal-to-distal* sequence which in effect produced an unstable motor strategy through a failure to control the joints closest to the weight-bearing

surface, allowing the distal segments to flail in an unstable manner relative to the supporting surface.

Such a reversal of motor patterning with brain damage indicates the importance of motor sequencing for normal motor control (see also the chapter by Woollacott). These findings illustrate how injury to the central nervous system may produce disordered motor control, but do not explore the role of muscles and their receptors in normal motor development and disordered states.

This chapter deals mainly with peripheral influences on motor control, it hopes to illustrate, mainly by reference to studies at the ankle joint, how apparently complex central phenomena may be influenced by relatively simple peripheral events and the significance of such a peripheral influence for the understanding of motor development and disordered motor control is explored.

The control of alternating movements will be described in terms of physical constraints, followed by a description of how the motor system is organised peripherally to meet these constraints. The concept of the functional joint range and optimum motorneurone output described in previous above (see sections 7,8), will be shown to play a crucial role in motor control.These findings provide a further scientific framework for the understanding of motor control and development as well as indicating how a variety of peripherally directed treatments might affect motor control.

9.2 The physics of motion and anatomical-inertial control.

i. All movements of the body are built up from the rotatory motion of limbs about joints, whether these are slow and smooth, or brisk and jerky. All movements involve overcoming inertia which is the property of all physical objects to resist a change in their state of rest or motion (Newton's First Law).

ii. Overcoming inertia requires force, which is the product of mass and acceleration (F=ma) and at their simplest level, all movements require a continuous matching of the forces developed by muscles with the inertial characteristics of the limbs.

iii Although the mass of the body remains invariable (ignoring the fluctuations in insensible losses), and the mass of the limb segments remains constant, the physiological rearrangement of the limb segments in relation to each other can profoundly affect the speed of rotatory movements by altering the inertial characteristics of the system. The maximum

speed of clapping hands with outstretched arms while restricting movement to the shoulder joints is both slower and more tiring than clapping from the wrist owing to the increased inertia of the outstretched arms compared with the hands alone. This is because the inertia of a limb can be shown to increase with the *fifth power* of the limb length, so that a doubling of limb length, as occurs in changing from a toddler to an adult, results in a 32-fold increase in the inertia (Walsh *et al*,1987), with obvious consequences for motor control (fig. 9.2). Relatively small peripheral changes can thus exert an enormous influence on motor speed: the commonest example being the spin of the ballet dancer or ice skater. Inertia is acceleration-dependent. It is the resistance to change in the state of motion or rest shared by all physical objects. For oscillating motion, the limb repeatedly changes direction, accelerating from a position of rest at each turning point. The frequency of voluntary alternating movements at a joint is therefore governed in absolute terms by the ability to overcome inertia, that is to say, the ability to modulate the force exerted by muscles.

iv. What are the physical laws governing alternating movements? For any oscillating system, the acceleration varies with the *square* of the frequency as given by the equation 1:

$$\text{Acceleration} = (2\pi\mathbf{f})^2\mathbf{A}\sin 2\pi\mathbf{ft} \qquad \textbf{equation 1.}$$

(**f**=frequency of oscillation, **A**=amplitude of oscillation and **t**=time)

According to equation 1, a doubling in frequency requires a *four-fold* increase and *trebling* of the frequency a *nine-fold* increase in the acceleration necessary to move the limb. Ultimately there are limits to the accelerations which muscles can generate which in turn limit the forces which they can develop.

v. The only way in which the frequency of oscillations can be increased is by diminishing the *amplitude* of oscillation. This is because the amplitude of oscillation of any system (including a limb about a joint) varies *inversely* with the square of the frequency of oscillation (equation 2) as can be seen by rearranging equation 1 so that for any given fixed maximum acceleration, the frequency of oscillation can only increase if the amplitude of movement is reduced:

$$\mathbf{A}=\text{Acceleration}/(2\pi\mathbf{f})^2\sin 2\pi\mathbf{ft} \qquad \textbf{equation 2.}$$

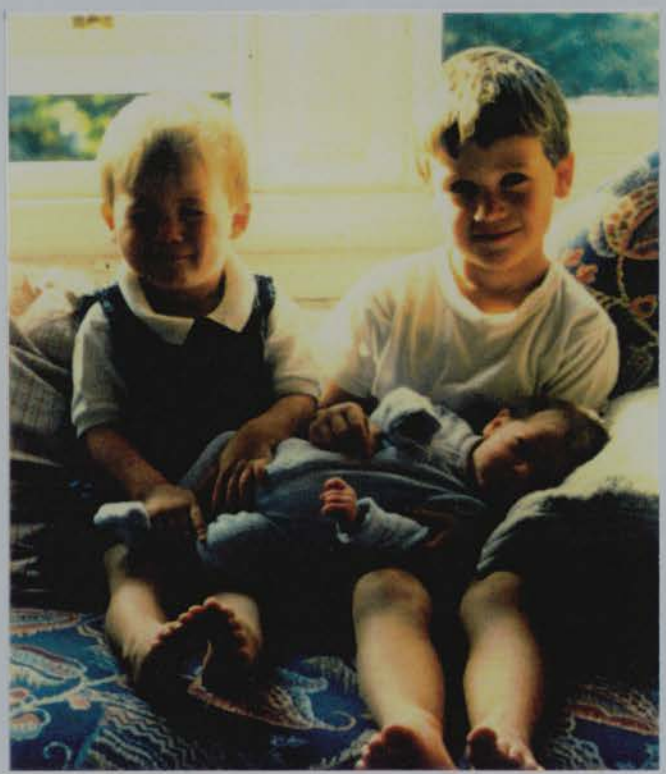


Figure 9.2 Limb length and limb inertia.
The long bones are more strikingly disparate in these three children aged approximately 2.5 years apart: new-born, 2.5 years, 4.5 years old. since the inertia increases with the fifth power of the limb length, each doubling results in a 32-fold increase in limb inertia (see Walsh and Wright, 1987).

It should be noted that equations 1 and 2 strictly refer to *linear* motion which is applicable to the intramuscular events. Similar expressions relating to *angular* motion exist in which angular velocity and acceleration replace linear velocity and acceleration respectively and mass is replaced by the moment of inertia. (see section 2.2.4, equations 6 and 7, above).

9.3 Amplitude and frequency of alternating movements.

The relationship between amplitude and frequency of oscillation is illustrated in figure 9.3.1 which shows voluntary isotonic alternating movements at the left ankle joint at increasing frequencies: as the frequency of voluntary oscillations increases, the amplitude diminishes. As can be seen the maximum angular velocities do not coincide with the fastest frequencies of voluntary alternating movements. These features are more clearly seen in figure 9.3.2 showing the first 65s from the same data. The first cursor is aligned with peak dorsiflexing velocity for large amplitude oscillations, which occurs at the mid range of the angular displacement, when angular acceleration is zero. The second cursor is aligned with peak plantarflexor velocity for large amplitude oscillations, again, occurring at the mid-range of the angular displacement. The third cursor is aligned with peak angular velocity at a higher frequency of oscillation, but note that the amplitude of motion has diminished: moreover, faster frequencies are achieved at lower amplitudes and lower angular velocities. It is also apparent from the traces for angular displacement shown with a compressed epoch in figure 9.3.1 and an expanded epoch in figure 9.3.2, that the maximum frequencies of oscillation are performed close to the neutral or "zero" angle, when the foot is at right angles to the shaft of the tibia. Also noteworthy is that the resting angle at the start of each voluntary motion sequence remains constant at -25° to -29° of plantarflexion beyond neutral. The subject begins by making large amplitude but low frequency movements which encompasses a joint range of 58°, with two thirds of the motion occurring in the plantarflexion range and one third of the motion dorsiflexed beyond neutral. At the fastest frequencies, the peak-peak motion amplitude has diminished to 12°, all of which is now dorsiflexed to just beyond neutral.

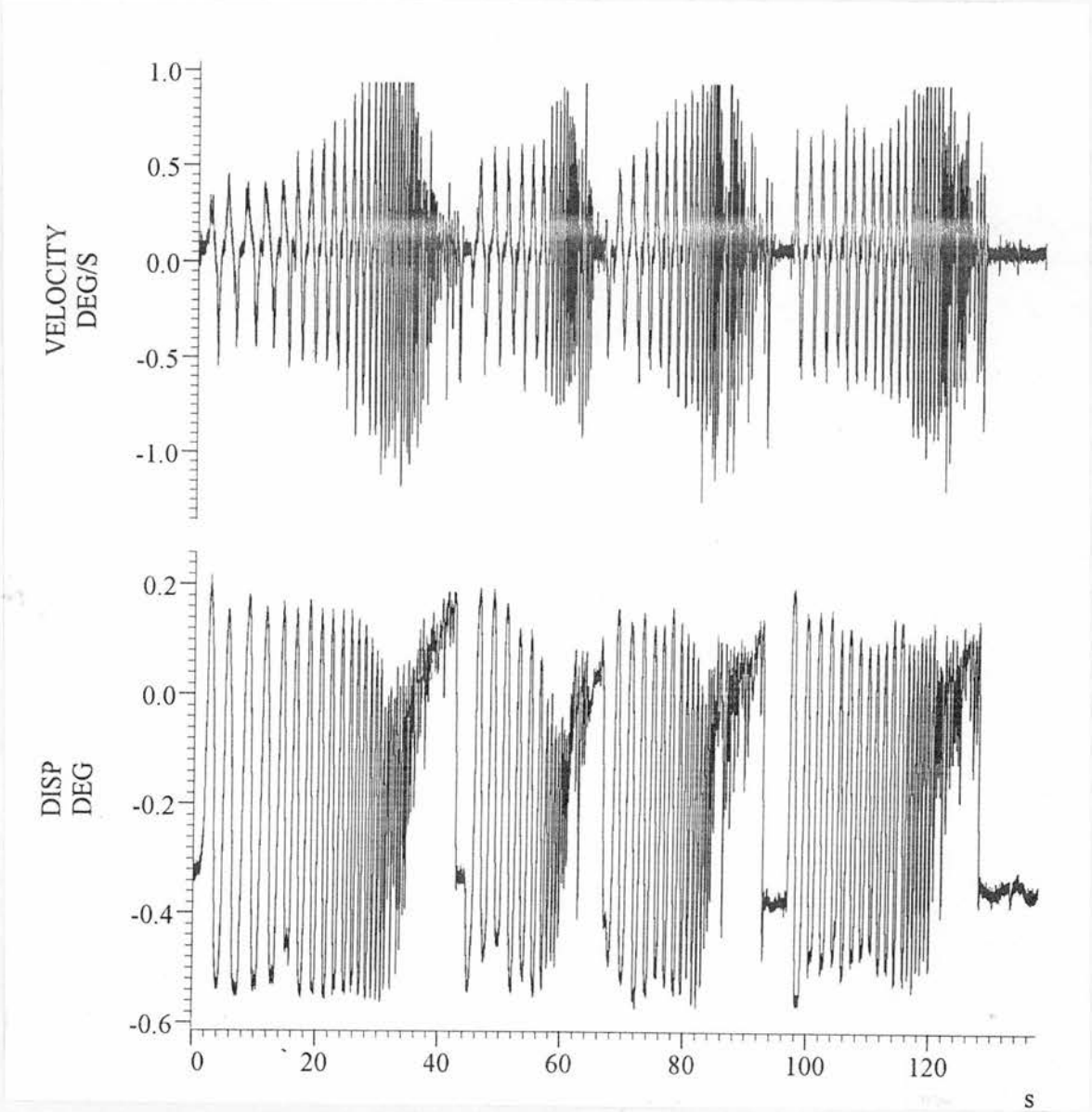


Figure 9.3.1 Voluntary alternating movements at the ankle joint.
Lt ankle, female, age 36 years, side-lying, gravity eliminated.
Above: angular velocity, 1 unit=80°/s.
Below: angular displacement, 1 unit=80°. 0°=neutral joint angle when ankle is neither plantarflexed nor dorsiflexed. Horizontal divisions=seconds. Four consecutive trials are shown, the subject being instructed to make the largest possible excursions to begin with and then to progressively increase the "speed" of alternating movements.Low-frequency movements are of large amplitude, but this diminishes as the frequency increases. In addition, the fastest frequencies are achieved either close to the neutral angle or in mild dorsiflexion beyond neutral. At the end of each trial, the foot returns to the position of resting plantarflexion which remains reasonably constant between -25° to -29°. The angular velocity saturates during the first, third and fourth trials. Nevertheless it can be seen that the peak-peak angular velocity reaches a maximum before the maximum alternating frequency is reached (see figure 9.3.2, cursor three, and figures 9.3.3 and 9.3.4)

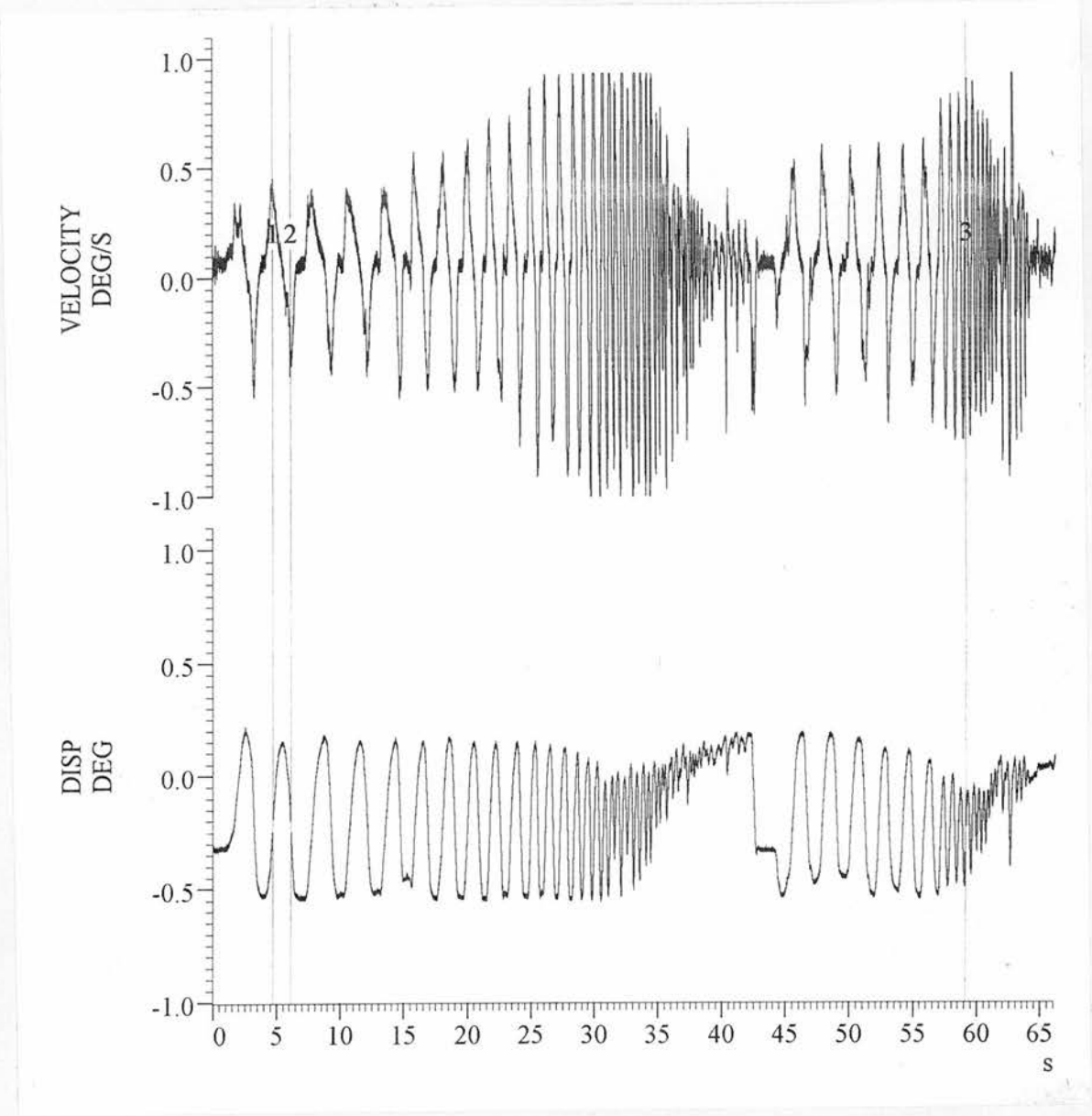


Figure 9.3.2 Angular velocity, displacement and frequency of alternating movements.
Lt ankle, female, age 36 years, side-lying, gravity eliminated.
Above: angular velocity, 1 unit=80°/s.
Below: angular displacement, 1 unit=80°. 0°=neutral joint angle when ankle is neither plantarflexed nor dorsiflexed. Horizontal divisions=seconds. Same data as the first 65s of figure 9.3.1. Cursors 1 and 2 are set at the peak dorsiflexing and plantarflexing angular velocities respectively, which coincide with the mid-range of the motion, when angular acceleration is zero. Cursor 3 is placed at the position of maximum peak-peak angular velocity occurs at about 2Hz at an amplitude of about 28° (see also figure 9.3.3). Faster voluntary frequencies are achieved at lower velocities and amplitudes. Note how in the first trial, the velocity trace saturates.

Figure 9.3.3 shows graphical plots of angular displacement and angular velocity against frequency of oscillation for the same limb and subject as in figures 9.3.1 and 9.3.2, as well as the data obtained from the right ankle. This right-handed subject achieves consistently higher frequencies for the right ankle, when compared with the left. For the right ankle and three consecutive trials, the peak angular velocity occurs at 2.7Hz, compared to about 2Hz for the left ankle. For both ankles, the maximum frequency of voluntary alternating movements occurs in the joint range just beyond neutral. Figure 9.3.4 shows data for voluntary alternating movements from right and left ankles of a child. The same features are evident as in the adult data in figures 9.3.1-3. In all the trials for right and left ankles, the maximum peak-peak amplitude is about 50° and the maximum alternating frequency of about 5Hz is in dorsiflexion relative to the resting joint angle and very close to the neutral joint angle. The peak angular velocity of just over 200°/s on the right and 154°/s on the left occurs at a frequency of 2Hz, the angular velocity declining as the frequency of oscillation reaches a maximum of about 5Hz. At a frequency of 1Hz, the amplitude of alternating movements is about 40°, declining to about 25° at 2Hz and 5° at 4Hz.

Similar results were obtained for three boys and two girls aged 8, 6, 3, 11 and 5 years.

9.4 Contractile properties of muscle and the functional joint range.

Simple, physical, non-neural constraints have been shown to regulate motion: whether a pendulum, a mass on a spring or bones, muscles and tendons at a joint, all systems have to overcome the same physical constraints. How have these physical constraints shaped the body's motor strategies? In isotonic alternating plantarflexion and dorsiflexion of the hindfoot at the ankle joint, slow oscillations can encompass the whole of the joint range but the fastest oscillations, which require the greatest accelerations (equation 1), seem to occur near the neutral angle at the ankle (figs. 9.3.1-4 and 9.4) ie they occur about the mid-range of the muscle's length. Such rapid oscillations are difficult or impossible at the extremes of the joint angle range unless very small amplitude movements are made.

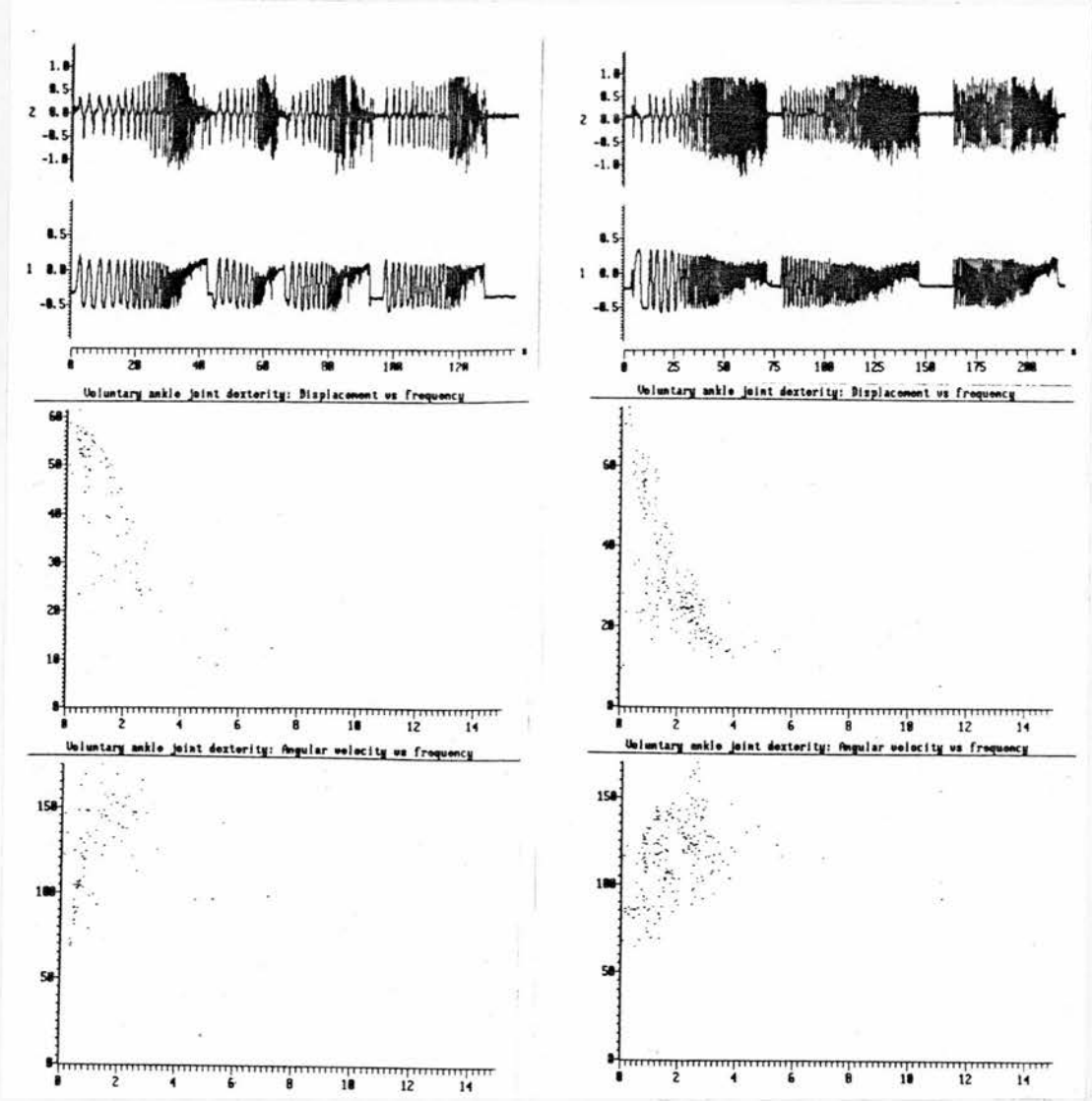


Figure 9.3.3
Angular displacement and velocity against frequency of voluntary alternating movements.
Lt and Rt ankle, female, age 36 years, side-lying, gravity eliminated.
Left hand figures: Lt ankle. Right hand figures: Rt ankle.
Top: traces of analogue angular velocity and angular displacement against time as for figures 9.3.1-2.
Middle: Angular displacement in degrees (vertical scale) against frequency of alternating movements (horizontal scale in Hz).The amplitude of motion declines as the frequency increases.
Bottom: Angular velocity in degrees/second (vertical scale) against frequency (horizontal scale in Hz). For the left ankle, the angular velocity reaches a peak of 158°/s at a frequency of 2Hz and amplitude of 28°. a slightly higher frequency of 2.7Hz is achieved on the right at a similar amplitude and angular velocity.
Lt ankle data as for figures 9.3.1-2.

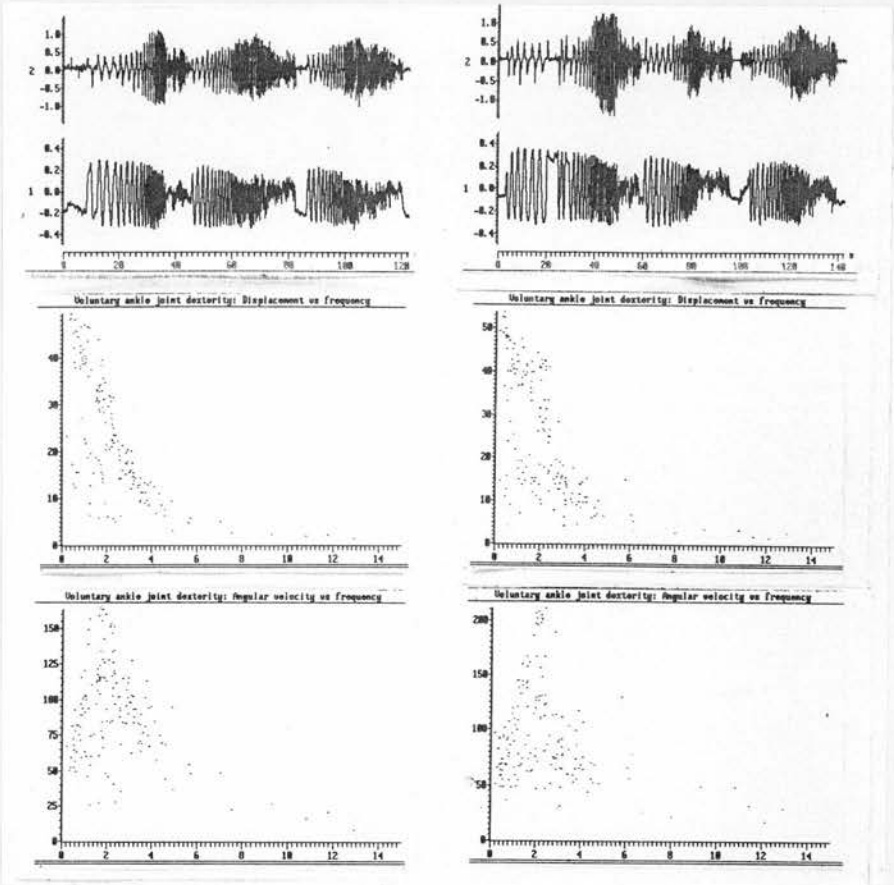


Figure 9.3.4
Angular displacement and velocity against frequency of voluntary alternating movements.
Lt and Rt ankle, female child, side-lying, gravity eliminated.
Left hand figures: Lt ankle. Right hand figures: Rt ankle.
Top: traces of analogue angular velocity and angular displacement against time.
Middle: Angular displacement in degrees (vertical scale) against frequency of alternating movements (horizontal scale in Hz).The amplitude of motion declines as the frequency increases.
Bottom: Angular velocity in degrees/second (vertical scale) against frequency (horizontal scale in Hz). The peak angular velocities for both ankles occurs at 2Hz at an angular displacement of about 30°, however the peak angular velocity on the right is 210°/s compared to 165°/s on the left. As in the other examples, angular displacement diminishes markedly with increasing frequency, to only a few degrees peak-peak at about 5Hz. The fastest voluntary oscillations are performed close to the neutral joint angle.

As has been demonstrated and discussed in the foregoing sections, the joint angle, both in isometric and isotonic conditions appears to modulate voluntary and reflex motor output (see sections 7.1 and 7.5). There appear to be at least three reasons in favour of an optimal neuro-mechanical angle relating to:

1. motorneurone recruitability, 2. muscle mechanical force and 3. temporal characteristics of contraction and relaxation.

1. As discussed in section 7. at the extremes of the joint angle, the muscle is weak, and only the weakest and slowest motor units can be readily recruited, whereas the largest and fastest motor units appear to be readily recruited (reflexly at least) at about-10° of plantarflexion.
2. The mechanics at the ankle or the arrangements of the calf muscle fibres or both are such, that the lever arm for the plantar flexors varies with the joint angle. So there are possible anatomical grounds for suspecting a functional joint angle
3. The temporal parameters are brief in the shortened state and up to 33% increased in the lengthened state, varying little from resting to the neutral angle.

Figure 9.4 illustrates the possible effects of a dysfunctional joint angle, ie posture on voluntary alternating movements at the ankle in a case of congenital left hemiparesis for boy with an equinus gait. With the subject side-lying to eliminate the influence of gravity, the nonparetic ankle movements conform to the patterns seen in healthy children and adults in figures 9.3.1-4, large amplitude movements begin at low frequencies of 0.5Hz between -66° and +26°. As the frequency of the voluntary movements increases so the the amplitude diminishes to about 13° peak-peak about neutral ie the zero angle. It is at this angle that the EMG discharges for the nonparetic limb achieve their highest amplitude and become most phasic, implying that voluntary recruitment of the largest and fastest (and most fatigable) motorunits occurs most readily at neutral.

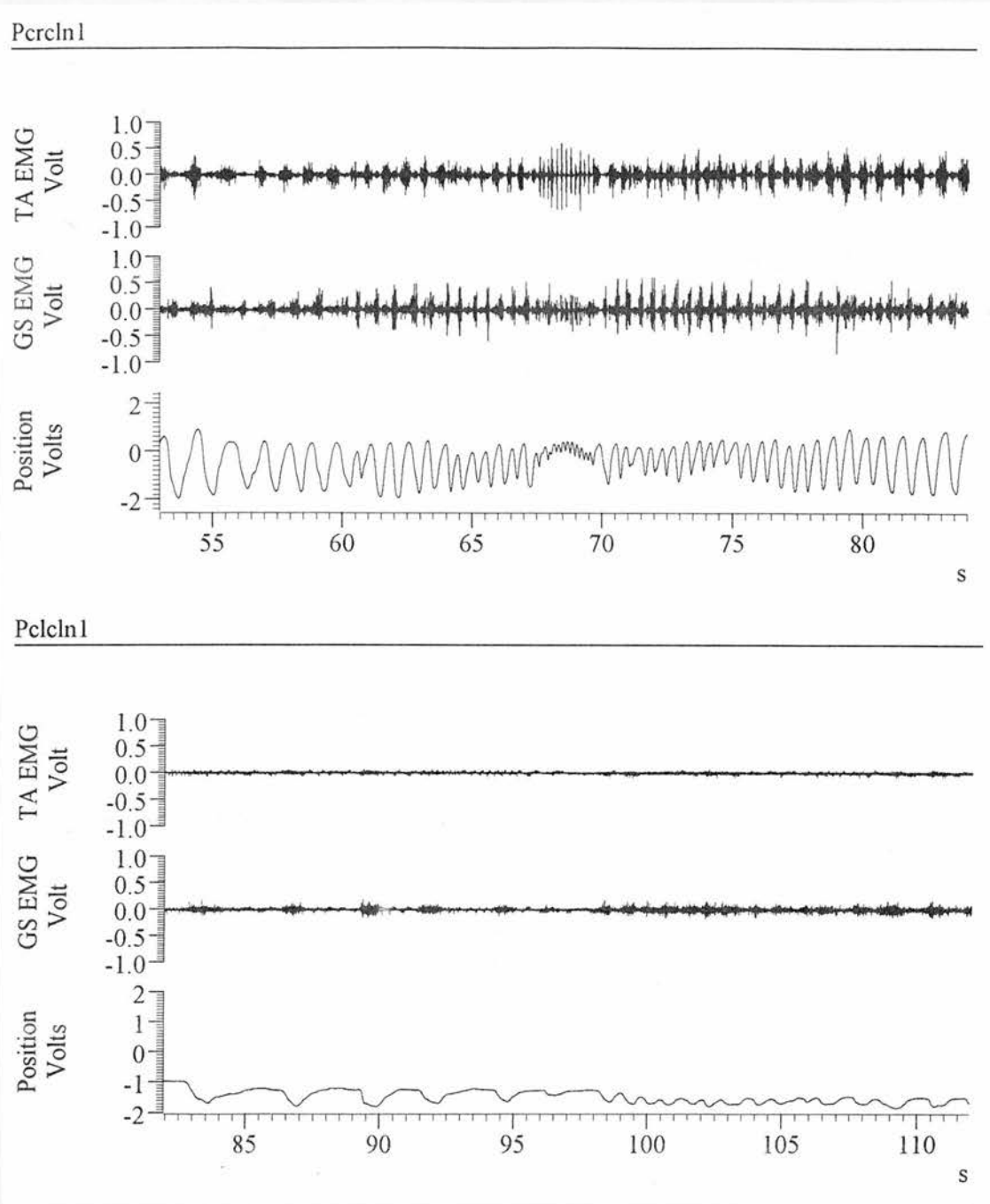


Figure 9.4 Alternating movements at the ankle in hemiplegia.
Top, non-paretic limb maximum frequency of voluntary movements occurs at low amplitudes near neutral angle, where the EMG discharges are highly phasic and of large amplitude, indicating recruitment of the fastest and strongest motor units.By equation 1, ow amplitudes allow the fastest movements.Bottom: hemiparetic limb: is in marked equinus. The EMG discharges are of low amplitude and poorly modulated and the the movements are correspondingly feeble.Horizontal divisions=0.5s, Vertical divisions 1 unit=33.25°, 0=neutral joint angle or right angle position of the foot with the shaft of the tibia. Negative values=plantarflexion, Positive values=dorsiflexion.

For the hemiparetic limbs, the resting joint angle is -33° of plantarflexion. Dorsiflexion beyond this appears impossible and the small amplitude, poorly differentiated movements of about 0.3Hz occur between -30° and -50° of plantarflexion, achieving a maximum frequency of 0.5Hz at a similar angle though with a correspondingly smaller amplitude of motion. The EMG discharges appear to be low and continuous for the hemiparetic tibialis anterior and gastroc-soleus muscles. One aspect of the disordered motor control appears to be diminishing dexterity partly arising out of a dysfunctional posture mitigating against favourable recruitment of spinal motor units

The *posture* of the limb thus seems to facilitate motor recruitment and output, ie it makes use of the peripheral neuromuscular constraints to modulate the desired action. One aspect of voluntary motor control may be to select the appropriate joint angle for the task, and during a continuously varying task, this changing joint angle contributes at a peripheral level, and at very short latencies to a continuous modulation of spinal motorneurone excitability, thus varying the motor output.

9.5 Cortical control of muscle force.

The foregoing sections have shown how the changing posture of the limb as it moves through a given joint range, continuously modulates motorneurone excitability.

Henneman and colleagues (1974) studied spinal motor unit firing in isometric tasks and demonstrated that a smooth increase in force output could be produced by a combination of first increasing the firing rate of motorunits and secondly by recruiting more motor units from a given spinal motorunit pool. Isometric tasks thus begin with the recruitment of slow and relatively weak (small) motorunits and as further force is required these spinal motorunits fire more rapidly until still further forces can only be achieved by recruiting larger, more rapidly firing motor units: the *Hennemann Size Principle* (Jones and Round, 1990).

At a cortical level, Evarts (1968) demonstrated that corticomotorneurone firing rate is correlated with force generation. These and other relevant studies are reviewed in depth by Porter and Lemon (1993, pp210-246). The modulation of the size and firing frequency of cortical and spinal motorneurones clearly determines the production of smooth and

sustained motor tasks.The relationship between motor unit recruitment and sensory feedback has been examined. It has been demonstrated that the highest sustainable firing rates of human motor neurones innervating intrinsic muscles of the hand are significantly lower in the absence of feedback from the target muscle (Gandevia *et al*,1990; Macefield *et al* ,1991) subjects were able to begin the task as well as perform ballistic movements but reported difficulties in sustaining a constant maximal effort as well as demonstrating evidence of reduced spinal motorneurone firing rates. So that although motor dexterity can under special conditions occur in the absence of feedback, it is obviously unnatural and difficult to sustain.

9.6 Alternating movements and age: do muscles develop?

When voluntary alternating movements (or oscillations) are measured at the ankle, wrist and metacarpophalangeal joints in children and adults (fig. 9.6), the frequency of such voluntary oscillations roughly doubles between the ages of three and eleven years, irrespective of the joint under study, and remains relatively constant thereafter (Lin, Brown and Walsh, 1996). Several factors could account for this speeding up, involving central and peripheral candidate structures.

9.6.1. Central candidate structures responsible for the increases in speed with age:

It is believed that unlearned, repetitive alternating movements in a single plane at one joint are generated in the primary motor strip of the pre-central gyrus(see Porter and Lemon 1993 for detailed discussion) in contrast to more complex tasks such as the orderly successive opposition of the finger tips with the thumb (as in the dexterity test of Martha Bridge Denckla, 1973) or even more complex opposition sequences which require increasing levels of concentration: these more complex movements appear to originate in the supplementary motor area as demonstrated by cerebral blood flow studies (Orgogozo and Larsen, 1979, Roland *et al* ,1980).

The supplementary motor area has an important role in the initiation and control of non-repetitive complex movements and Orgogozo and Larsen refer to it as a "supramotor area" which is functionally of a higher hierarchical order than the primary Rolandic areas. Activation of the supplementary motor area has been demonstrated noninvasively by functional magnetic resonance imaging in healthy adult controls (Santosh, Rimmington and Best, 1995.

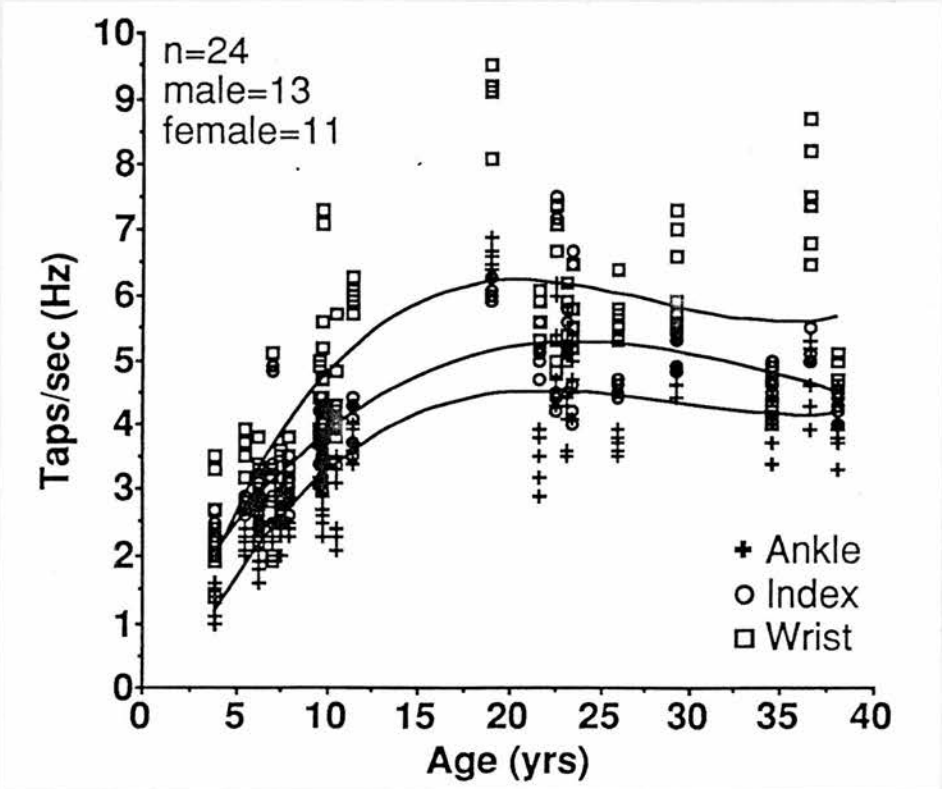


Figure 9.6
Frequency of alternating movements at the ankle,metacarpophalangeal and wrist joints
Ages 3 to 70 years.There is a doubling of the voluntary alternating movements in the first decade of life.The fastest movements occur at the wrist, followed by metacarpophalangeal and the ankle joint. *After Lin, Brown and Walsh, 1996, by permission .*

Work on the cerebral localisation of complex motor tasks demonstrates the complexity of the cerebral organisation of movement and a number of studies have shown that a variety of surgical lesions can impair alternating movements. Ablation of the medial part of the frontal lobe which includes the supplementary motor area in three patients resulted in a characteristic sequence of recovery:

stage 1 a post operative global akinesia which was most prominent contralateraly and associated with speech arrest,

stage 2 a spontaneous motor recovery albeit with a marked reduction in spontaneous movements, facial expression and speech

stage 3 a disturbance of alternating movements of the hands long after the other functions had recovered (Laplane *et al* 1977).

In another study, stimulation of the "frontal speech area" during the course of epilepsy surgery investigations resulted in combinations of speech arrest, writing arrest or impaired rapid alternating alternating movements of the tongue, fingers and toes (Lesser *et al*, 1984). These authors used chronic subdural electrode stimulation techniques to demonstrate that rapid finger and tongue movements could be interrupted by stimulating over quite wide areas of the posterior aspects of the third frontal gyrus.

It is reasonable to suppose that in healthy development, there is a maturation of neurones and their connections involved in motor tasks.Likewise, it is possible to demonstrate that as a child's walking matures from infancy, so that cocontractions of muscle groups which produce stiff and ungainly movements, give way to a phasic interaction of EMG activity leading to joint asynchronies and smoother movements (Leonard *et al* 1991, Sutherland *et al*, 1988).

This maturation suggests an emerging motor engram (both spinal and cortical) for walking and other movements. Notwithstanding central nervous maturation it is worth pausing to consider other factors. In this context, the observations of Denckla (1973) demonstrating a speeding up of sequential finger movements with age in children has been attributed almost entirely to the maturation of central structures, although a peripheral contribution to this maturation is described below.

9.6.2 Corticospinal tract maturation.

Central conduction from the corticomotorneurone to the spinal motor neurone has

been shown, using magnetic stimulation techniques, to reach a maximum speed by 18 months of age (Eyre *et al*, 1991), (although another study, also using magnetic stimulation techniques has suggested a longer period of maturation of 8-10 years see below), Eyre and colleagues conclude:

" From the age age of 2 years, the fastest conducting fibres in sensory and motor central pathways operate with constant conduction delays"

Eyre, Miller and Ramesh, 1991.

It is thus unlikely that central conduction delays are responsible for the increasing speed of alternating movements between 3 and 11 years of age, however, this is disputed by Müller and Homberg (1992) who also demonstrated that the fastest repetitive movements reach a maximum at the end of the first decade of life. In contrast to Eyre and colleagues, Müller and Homberg attribute this increase in speed to a reduction in central conduction latency over the first decade.

9.6.3 Peripheral candidate structures for the maturation of dexterity

Possible peripheral structures that might account for the increases in speed with age include:

i. Peripheral nerve conduction velocity, which more than doubles from birth to adulthood from 28.5 m/s to 82 m/s. Despite this, the latency (conduction time) along the nerves increases because of their increase in length so that the latency of the H-reflex latency (see section 7), increases from 15 to 28ms between birth and adulthood (Mayer and Mosser, 1973). Such an increase in nerve conduction latency would have a tendency to slow down the speed of alternating movements with age, and be at variance with the observed speeding up.

ii. Muscles themselves could provide the rate-limiting step. The changes with age of reflex twitch force and soleus twitch frequency (the reciprocal of the soleus twitch time) are illustrated by figure 9.6.3.1 for males aged 3.9, 10.5 and 19 years. The soleus twitch force clearly increases with age. Likewise the isometric soleus twitch frequency at neutral increases from 3 Hz at 3 years and 4.5 Hz at 10.5 years to 5Hz at 19 years. Similar changes with age are measurable in females, the soleus twitch time at neutral increasing from 2.5Hz at 3 years to just over 4Hz in the early 20s.

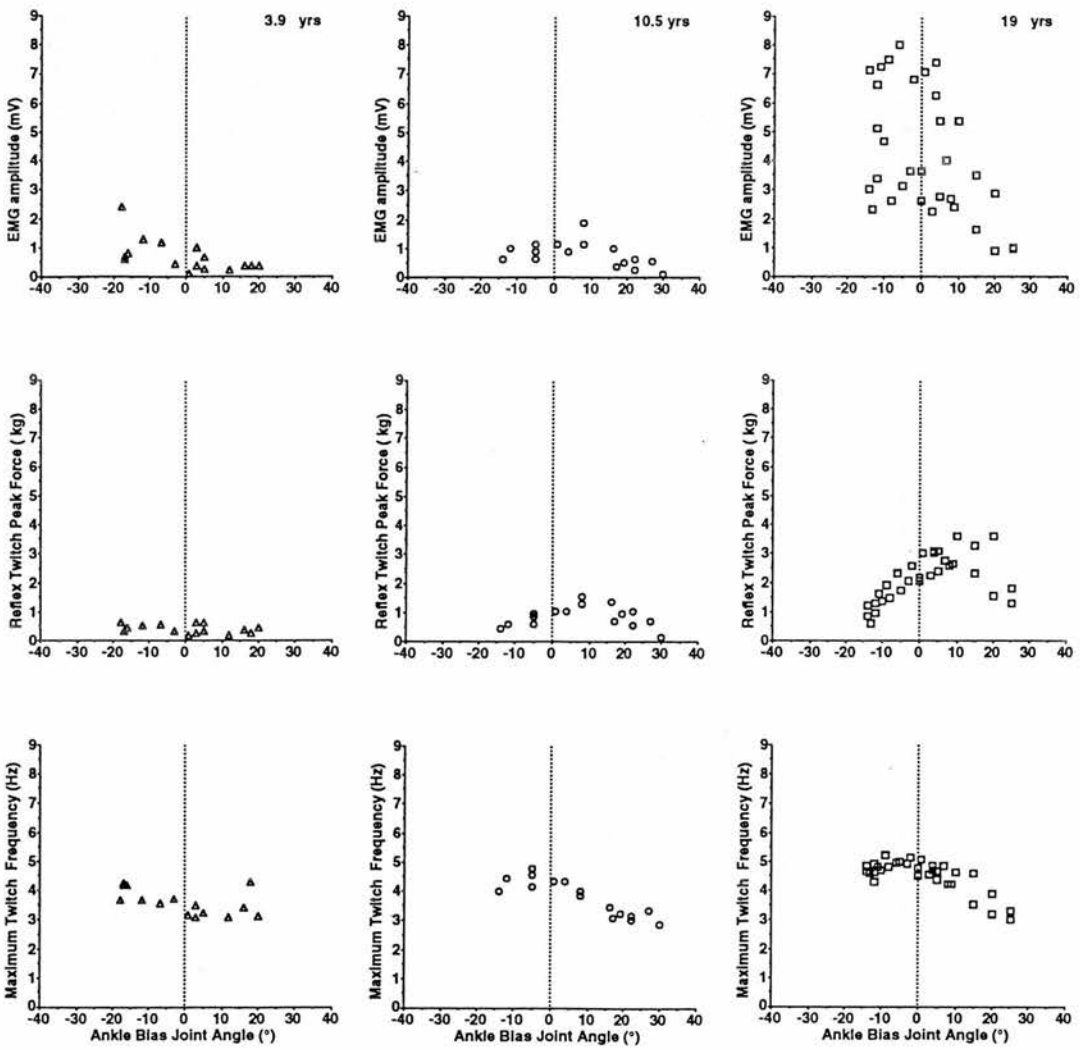


Figure 9.6.3.1 Maturation in soleus muscle reflex twitches.
Top row : Reflex EMG amplitude (mV). Middle row: Peak reflex twitch force (N), bottom row: Maximum twitch frequency. From left to right: boy aged 3.9 years, boy aged 10.5 year and an adult male aged 19 years. Twitch force obviously rises from about 2.45-4.9N at age 3 years to 29.4-39.2N in adulthood. Likewise the twitch frequency rises from 3Hz at the neutral angle at age 3-4 years increasing in frequency to 5Hz at age 19 years.
NB: Each point represents an average of four consecutive taps.

After Lin , 1997

Scott and colleagues (1985) found it difficult to fatigue the tibialis anterior muscles of healthy children implying that children exhibit slower, non-fatigable, type 1, muscle phenotypes with an oxidative metabolism. Brooke and Engel (1969) showed that muscle fibre diameter increases with age so that sex differences begin to emerge round about 10 years of age. There is some histochemical evidence to support the notion of muscular maturation with age although the two most recent histological studies offer conflicting evidence.

A maturation from a slow to fast phenotype was demonstrated by Lexell and colleagues (1992) in a study involving 22 muscle biopsies from subjects aged 5 to 36 years who had died accidentally: a slow muscle phenotype of the vastus lateralis muscle being clearly evident in the children. Elder and colleagues (1993) demonstrated in 19 newborns and 35 children that while the tibialis anterior muscle half-relaxation time speeds up between 5-16 months of age, that of the posterior calf muscles slows over a similar period. Nevertheless, some infants demonstrated persistence of slow muscle phenotypes and in a parallel study of 43 subjects aged 22 weeks gestation to 28 years, there was an overall preponderance of slow (type 1) muscle phenotypes in children as compared to adults from samples of triceps brachii, vastus lateralis, biceps brachii and tibialis anterior muscles.

Some of the mechanisms by which the twitch profile might speed up with age have been studied (Margareth and colleagues, 1980; Zubrzycka-Gaarn and Sarzala, 1980) and may relate to the ontogeny of the sarcoplasmic reticulum enzyme biochemistry which regulates calcium re-uptake and hence muscle relaxation. It is not known what factors may influence such a muscle maturation although a variety of internal and external stimuli are known to affect the myosin-ATPase expression in muscle. More recently, variations in myosin isoforms with age have been identified using immunocytochemical techniques capable of detecting myosin heavy chains. This has been reviewed by Whalen (1985):

"The scenario that has emerged concerning the major myosin isoenzymes in skeletal muscle is the following. Soon after their formation, muscle fibres contain an embryonic (or foetal type of myosin, which is subsequently replaced by a neonatal isoenzyme. Eventually (e.g. 3-4 weeks after birth in rats and humans), adult myosins become established as the predominant species and they persist into adulthood."

Whalen (1985), p 45.

"The adult forms are responsible for determining, at least in part, the performance of muscle tissue, while other forms are characteristic of developing muscle and possibly provide some special-as yet undiscovered-function for these muscles. These developmental isoforms are also reexpressed in regenerating muscle (Sartore, Gorza and Schiaffino, 1982; Maréchal, Schwartz, Beckers-Bleukx and Ghins, 1984) and regeneration is a feature of several muscle diseases (Fitzsimons and Hoh, 1981a). Finally, there is evidence that aberrant expression of the developmental forms occurs in some physiological situations independent of regeneration (Fitzsimons and Hoh, 1981b).

Because of the association of certain myosin isoenzymes with different physiological or pathological states of muscle, the transitions between different isoenzymes and the control of these transitions are worthy of careful study. Since most physiological and developmental studies concern the hind limb muscles, current efforts normally take into account the four major forms found in these muscles: embryonic, neonatal, slow and fast."

Whalen (1985), p 47-48

In developing rats, that mature in the first 3-4 postnatal weeks, neonatal and adult myosins coexist over several weeks in the same fibres and the ontogeny of fast myosin isoforms appears independent of innervation (Whalen, 1985; Butler-Brown *et al*, 1982). Whalen (1985) points to the natural progression for muscles to progressively change from expressing embryonic-neonatal-fast myosins over time, though these isoforms may convert to a slow isoform at any stage, a process which may be nerve-driven.

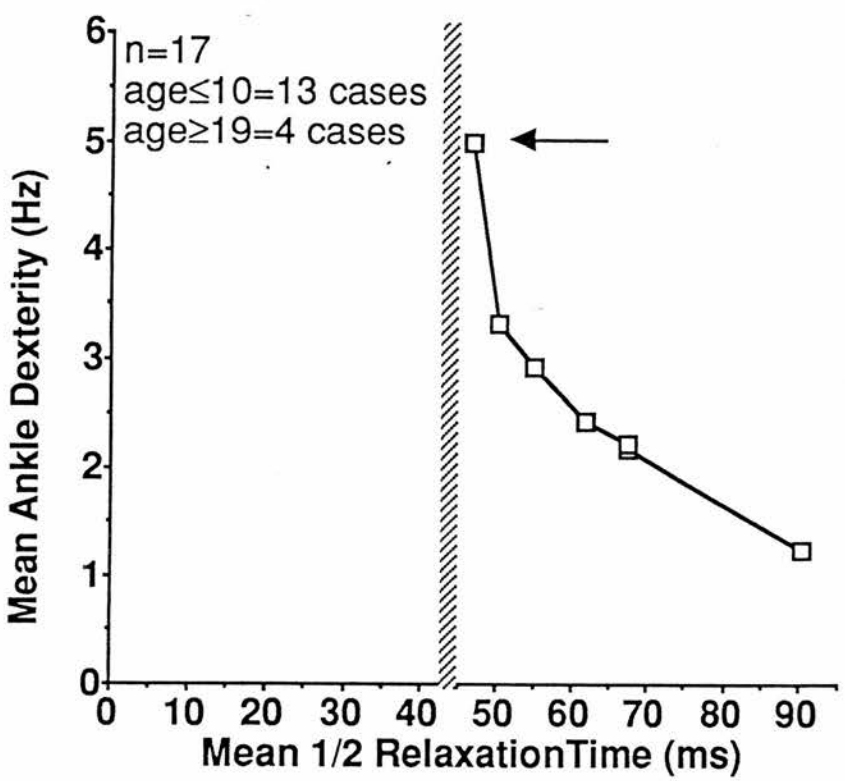


Figure 9.6.3.2 Ankle dexterity frequency and soleus muscle twitch 1/2 relaxation time.
The mean 1/2 relaxation time for young adult males is 43.8+/- 10.7 ms and 56.8+/- 12.2ms for young adult women (Walsh *et al*, ,1992).*Fom Lin Brown and Walsh, 1996, by permission* .

"The denervation experiment described above suggests that the embryonic-neonatal-adult fast pathway is nerve-independent, while considerable evidence from other experimental models (reviewed by Jolesz and Sreter, 1981) strongly suggests that slow myosin accumulation is a nerve-driven process, at least in some adult animals."

Whalen (1985), p 49.

Another regulatory factor is thyroid status:

"While the results of denervation experiments have suggested that the neonatal-adult fast transition is nerve-independent, recent investigations have revealed that it might be critically regulated by thyroid hormone levels. If rats are treated with antithyroid drugs during gestation and into the first month after birth, the hypothyroid offspring will accumulate neonatal but not adult fast myosin (Gambke et al, 1983, Butler-Browne, Herlicoviez and Whalen, 1984)."

Whalen (1985), p 49-50.

That muscle properties change from the myotube of the foetus to the adult is not in doubt although it is advanced in this chapter,that the time scale for such muscles change is prolonged well beyond the first few years of life and certainly involves more than a change in the fibre diameter and muscle compliance: changes in the myosin isoforms may occur more slowly in children to account for the prolonged immaturity of the motor system in humans.

The notion of muscle time constants is beyond the scope of this work but has allowed for some interesting observations in comparative anatomy as between large and small mammals:

" An apparent paradox is also found in animals of different size. The time constants of of contraction are determined by myosin isoforms. It has been shown that the molecular structures of slow and fast myosin are very similar throughout mammalian muscles. The time constants are, however, also affected by body size such that they decrease as body mass increases, in proportion to $M_b^{-0.25}$. This leads to the paradox that slow muscles of the mouse are actually faster than the fast muscles of the cat-and still the cat catches the mouse!"

Weibel, 1985, p409-410.

Evidence presented in section 7 (fig. 7.4.5.1d) of this thesis, is that young human muscles are intrinsically slower than adult muscles, even though young humans are smaller than adults!

At least three principle mechanisms for the observed increases in motor speed in the first decade of life can be identified. The first of these is the undoubted increase in force generation crucial for overcoming inertia and hence the production of faster movements (equation 1, above) with age. This can be demonstrated at the level of isometric reflex twitches (see section 7.4.6.3, fig. 7.4.5.1c). Secondly, faster muscle relaxation times should facilitate the production of faster alternating movements. Thirdly, there may be a learning curve for the performance of rapid tasks involving experience of the functional joint range and optimal joint angle.

9.7.1 Muscle speed, joint angle and the secondary effects of central motor impairment.

A corollary to these properties of muscles is that in the presence of a central motor disorder, fibre-type reverts to a slow isoform (Edström, 1970;Young and Mayer, 1982; Dietz *et al*, 1985). In addition to this, motor unit numbers diminish by about 50% while the size of each motor unit increases (McComas *et al*, 1973) within a year after a stroke: an effect that would influence motor unit recruitment in voluntary tasks. Added to this, secondary atrophy and deformities through disuse and the dysfunctional joint angles and it becomes clear how the primary, central motor disability is exacerbated

by peripheral changes to the neuromuscular system (figure 9.4).

9.7.2 Muscle speed, joint angle and the ontogeny of alternating movements.

What is the practical significance of these observations for child development? Evidence for motor maturation can also be seen in pictures, for instance where colouring in shapes is required, a task which requires alternating movements at the wrist and forearm. The 4 year old makes broad, coarse (slow) strokes (fig 9.7.2, top) whereas the 7 year old uses short, close-knit (rapid) strokes which provide a denser, more even covering of the paper (fig 9.7.2, lower). In their every-day lives, children's' movements show an adaptation to the physical and neuromuscular constraints described above. It is hypothesised that the older child optimises the neuromuscular output by appropriate postural adjustments which include:

i. Making small amplitude strokes which in turn allow a higher frequency of alternating movements and denser colouring strokes.

ii It is also likely that the older child's rapid strokes are being performed (unconsciously) close to the functional mid-range of the muscle for optimal neuro-mechanical coupling.

The younger child, by making large amplitude strokes, will produce slower movements since he/she is working quite literally at a physical disadvantage.

Despite these compelling arguments for a peripheral control of motor function by means of short-loop spinal modulation presented here and by others (Hagbarth, Wallin and Löfstedt, 1975; Al Falahe, Nagakoa and Vallbo, 1990, Gandevia *et al* ,1990; Macefield *et al* ,1991) there are isolated case reports of voluntary alternating movements occurring in the presence of complete deafferentation in man (Rothwell *et al* ,1982) and in primates (Polit and Bizzi, 1979). The extent to which such movements can be modulated remains unclear. Also,

these findings were obtained in adults who presumably had experienced normal sensory feedback as children (or young primates) so that the situation in these cases is of a loss of a sensory modality rather than development from birth without sensory feedback. These interesting questions are dealt with in some depth by Jeannerod (1988 pp 171-208).

Figure 9.7.2 Colouring skills in children

See Overleaf.

The ability to rapidly colour in may reflect the changes in motor control described in this chapter. **a.** 4 year old boy shading: the coarse (slow) strokes of large amplitude are unable to adequately fill the geometric shape. **b**. 7 year old boy: fine, closely packed (fast) , short strokes of colouring which completely fills in the shape.

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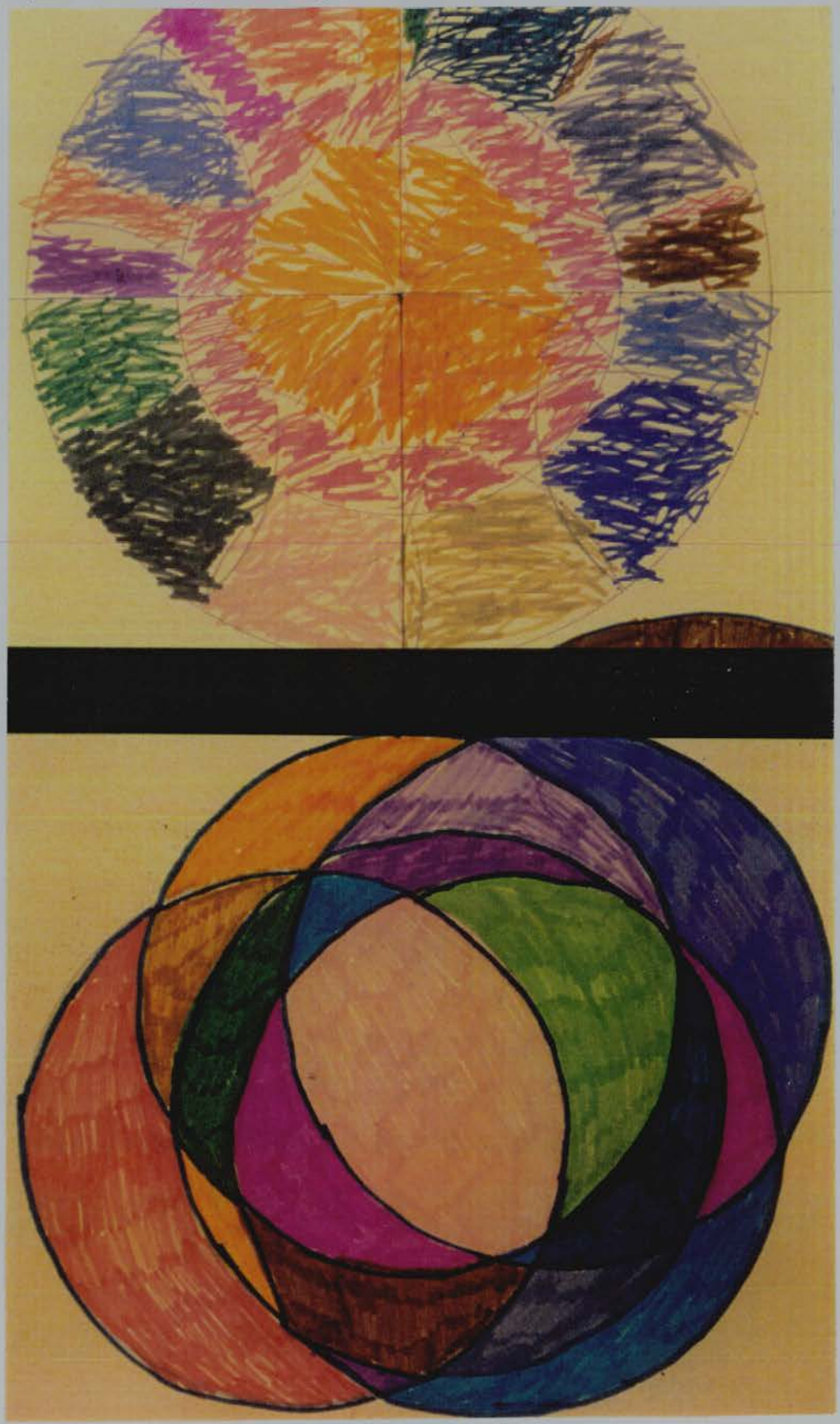

Figure 9.7.

9.8. Acknowledgements.

Much of the material contained in this chapter has been published elsewhere in joint collaboration with Dr JK Brown and Dr EG Walsh in: Lin, Brown and Walsh, 1994; 1996, 1997 and Lin, 1997 and the relevant papers are appended.

10. Posturing in cerebral palsy

10.1 Background.

The distinction between posture and movement is not easy, since they are invariably interdependent. Nevertheless, it can be inferred from observation that motion occurs with respect to the background attitude of the other body parts.

Abnormalities in posture have been discussed and illustrated in several parts of this work together with emergent or developing postures in relation to standing, walking (sections 1-4) and running; and as a contribution to muscle tone, or the resistance felt when stretching muscles (section 2). Particularly in section 3, attention has been drawn to a variety of mechanisms likely to contribute to the common problem of equinus with the conclusion that equinus is not so much a product of muscle weakness or of imbalance of power across the ankle joint, nor is the dominant feature reflex excitability. Instead, more emphasis was placed on developmental immaturity, the release of centrally determined postures and abnormal patterns of muscle activation

In section 4, attention was drawn to non-electrical changes in the plastic muscle properties which could be exploited to allow the foot to adopt a new posture for several minutes after slow passive stretching: again, a phenomenon not maintained by abnormal stretch reflexes. In sections 5 and 6, muscle length was shown to influence reflex excitability, while in sections 7 and 8, the relationship between muscle length, reflex excitability and muscle twitch characteristics is explored in greater detail. In all of these studies, the posture (muscle length) exerted a primary influence on the phenomena (stretch reflexes and the muscle twitch *per se*). The possible role of posture in regulating motorneurone output at moderate levels of activity has been advanced in section 9, along with the suggestion that children learn to optimise posture (joint angle) to produce increasingly efficient movements.

The purpose of this section is to briefly mention tonic neck reflexes, tonic labyrinthine reflexes and Føg's posturing all of which appear to be influences on posture in varying degrees during normal development, and particularly after brain injury. In the case of Føg's posturing, unusual postures emerge in perfectly healthy children during certain tasks, but may become an obligate accompaniment to movement after brain injury.

10.2 Tonic neck and labyrinthine reflexes: basic review of function.

The phenomena of tonic neck and tonic labyrinthine reflexes has been extensively

reviewed by Magnus and de Kleijn (1912) in the cat, Walshe (1923) following hemiplegia, Roberts (1968) in different quadrupeds and man, Chan (1983) in man, with particular reference to otolithic influences on lower limb motorneurones and the H-reflex, and by Brown (1997) in the developing foetus, infant and in abnormal motor states.

The findings of Magnus and de Kleijn (1912) were summarised by Walshe (1923):

"The researches of Magnus and his school have been devoted to these regulating mechanisms and may now be said to have solved the main problems in the co-ordination of posture. Beginning with the decerebrate animal it was found that the otolith organs of the labyrinths (and not the semicircular canals) and the proprioceptors of the neck are the sources of impulses which by their influence on muscle tone produce reflexly a whole variety of attitudes of the limbs and the trunk. Thus, changes in the position of the head (otoliths) in relation to the horizontal plane of space cause an identical variation in the extensor tonus of all four limbs. The maximal influence is exerted when the animal is inverted, and the minimal when the animal stands on its feet in a natural posture. Alterations in posture of the head in relation to the trunk also produce variations in limb tonus and therefore in attitude, quite apart from labyrinthine influences. Thus, rotation of the head to one side causes increase of extensor tonus in the limbs on the side to which the snout is directed, and diminution of extensor tone in the crossed limbs. Retraction of the head produces extensor spasm in the fore limbs and loss of extensor tone in the hind limbs, and ventral flexion of the head has the opposite result "

FMR Walshe, 1923.

The combined influences of tonic labyrinthine and tonic neck reflexes has been described by Roberts (1968), in which nine different basic limb postures can be produced according to the combinations of 'head up', 'head normal', 'head down' with 'neck dorsiflexed', 'neck normal' and 'neck ventriflexed'. These combinations can be interpreted as functional according to terrain and direction of motion, for example standing on an upward or downward slope, looking up, looking ahead, looking down. Under certain conditions, the labyrinthine and tonic neck reflexes cancel each other out. Normally, 'head up' produces foreleg flexion and hindleg extension, whereas neck dorsiflexed produces foreleg extension and hindleg flexion: if 'head up' and 'neck up' are combined, forelegs and hind legs remain unchanged. (top left panel, fig. 10.2).Similarly, if 'head down', which usually produces foreleg extension and hind leg flexion, is combined with 'neck down', which usually results in foreleg flexion and hind leg extension, then again the limbs remain unchanged (fig. 10.2, bottom right panel). If the the head is in the plane of the body (head normal) and the neck is neither flexed nor extended (neck normal), once again the limb posture remains unchanged (fig. 10.2, middle panel). It therefore seems that if the head and neck tend to the same posture, there is no tonic influence on the limbs.With the head in plane with the trunk (neck normal), but the animal going uphill (head up) the forelegs are flexed and hind legs extended (fig.

10.2, middle left panel). If the head is in plane with the trunk (neck normal) and the animal is going downhill (head down) the forelegs are extended and hind legs flexed (fig. 10.2, right panel).

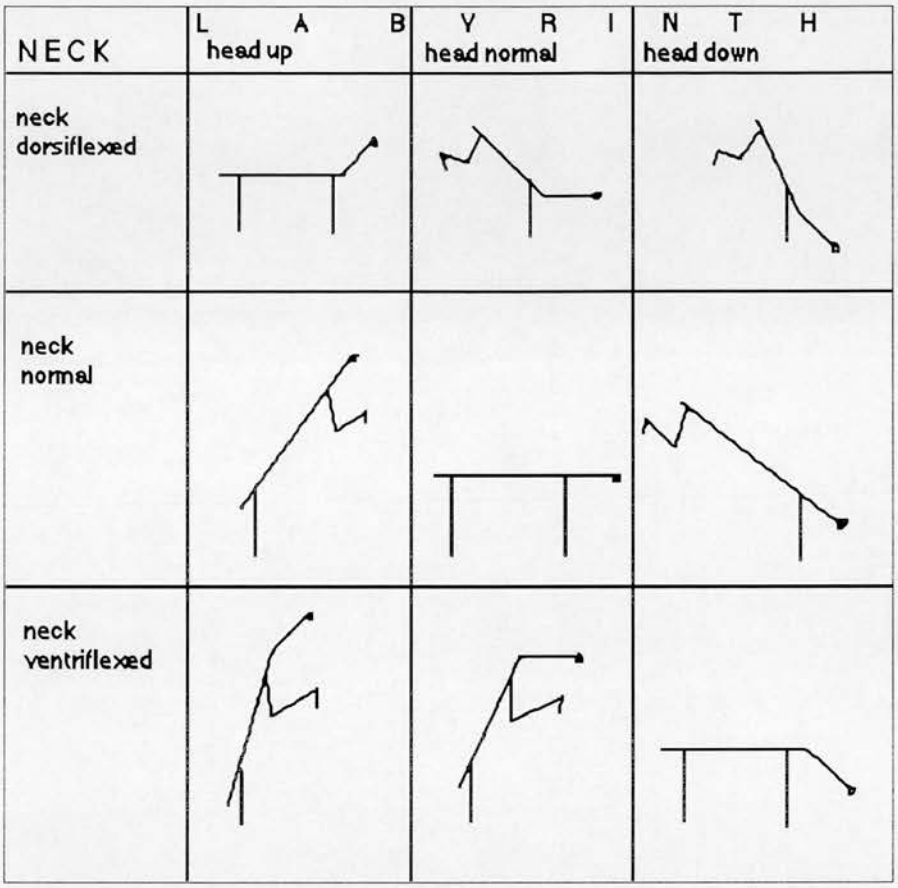

Figure 10.2 Interaction of tonic neck reflexes with tonic labyrinthine reflexes
Adapted from Chan, 1983: after Roberts, 1968.

Finally, 'head down' with 'neck dorsiflexed' facilitates forelimb extension and hind-limb flexion (fig. 10.2, top right), whereas, 'head up' and 'neck ventriflexed' produces the reverse (fig. 10.2, bottom left).

The interaction between the tonic neck and labyrinthine reflexes are clearly complex, and the number of possible subtle permutations of combinations between the two are well-nigh infinite.

Persistent tonic neck and labyrinthine reflexes after infancy is a hall-mark of neurological dysfunction commonly encountered in paediatric practice and has been referred to as a poor prognostic indicator for independent walking in the section on the natural history of cerebral palsy (section, 1.3.4). Denny-Brown (1980) comments extensively

on the relationship between cerebral lesions and tonic labyrinthine postures:

"In our own studies of the effect of cortical ablation in the monkey, we found that the spasticity reported by Fulton and Kennard, resulting from combined ablation of areas 6 and 4, is in fact quite mild in degree, though the upper limb is held slightly flexed and the lower slightly extended, with corresponding increase and spread of tendon reflexes. This posture is reversed if the animal is held downward. The spasticity and reaction to inversion are much more intense if the the cortical ablation is performed bilaterally. Ablation of area 4 alone (Denny-Brown and Botterell, 1947) led to increase in spread of tendon reflexes in the fingers and wrist. If the limb was deafferented, the flexed posture of the upper limb and its reversal by inverting the animal persisted (Denny-Brown, 1966). The tendon reflexes were of course abolished by such deafferentation. "

Denny-Brown, 1980, p6

The important observation which Denny-Brown makes is that the posture of the animal is not altered by deafferentation although the tendon reflexes are abolished. He emphasises the need to distinguish between the spasticity, ie the velocity-dependent increase in stretch reflexes which is dependent, by definition on stretch-receptor afferent input, and the posture which is independent of these velocity-dependent reflexes.

The release of extrapyramidal phenomena with more extensive lesions is referred to in the same passage:

"It was also found that if, in addition to areas 6 and 8, area 7 in parietal cortex with a related strip of parietal and frontal operculum was removed, leaving pre-and post-central gyrus intact, a diffuse rigidity with emphasis on flexor muscles appeared (Denny-Brown, 1966). Removal of this 'perirolandic cortex on both sides created an extrapyramidal rigidity with prominent release of labyrinthine and neck reflexes. The extrapyramidal effects of removal of precentral cortex described by Fulton and Kennard appear to be just a fraction of this type of extrapyramidal pattern. The classical prototype of spastic hemiplegia familiar to physicians as e result of lesions in the internal capsule now appear to be the result of loss of both pyramidal and extrapyramidal descending motor pathways"

Denny-Brown, 1980, p6

Here, Denny-Brown (1980) makes important observations in relation to the tonic neck and labyrinthine reflexes as extrapyramidal manifestations of perirolandic lesions, with the corollary that the presence of tonic neck and labyrinthine reflexes denotes an extrapyramidal phenomenon. His second important observation is to describe the mixed pyramidal and extrapyramidal effects of a capsular lesion.

10.3. Tonic labyrinthine reflexes in diplegia of prematurity.

The following demonstration has been reproduced repeatedly during the course of clinically examining children with diplegia. In this instance, the variations in posture with position of the head in space is accompanied by surface EMG recordings of the thigh muscles. Figure 10.3 illustrates the salient features.

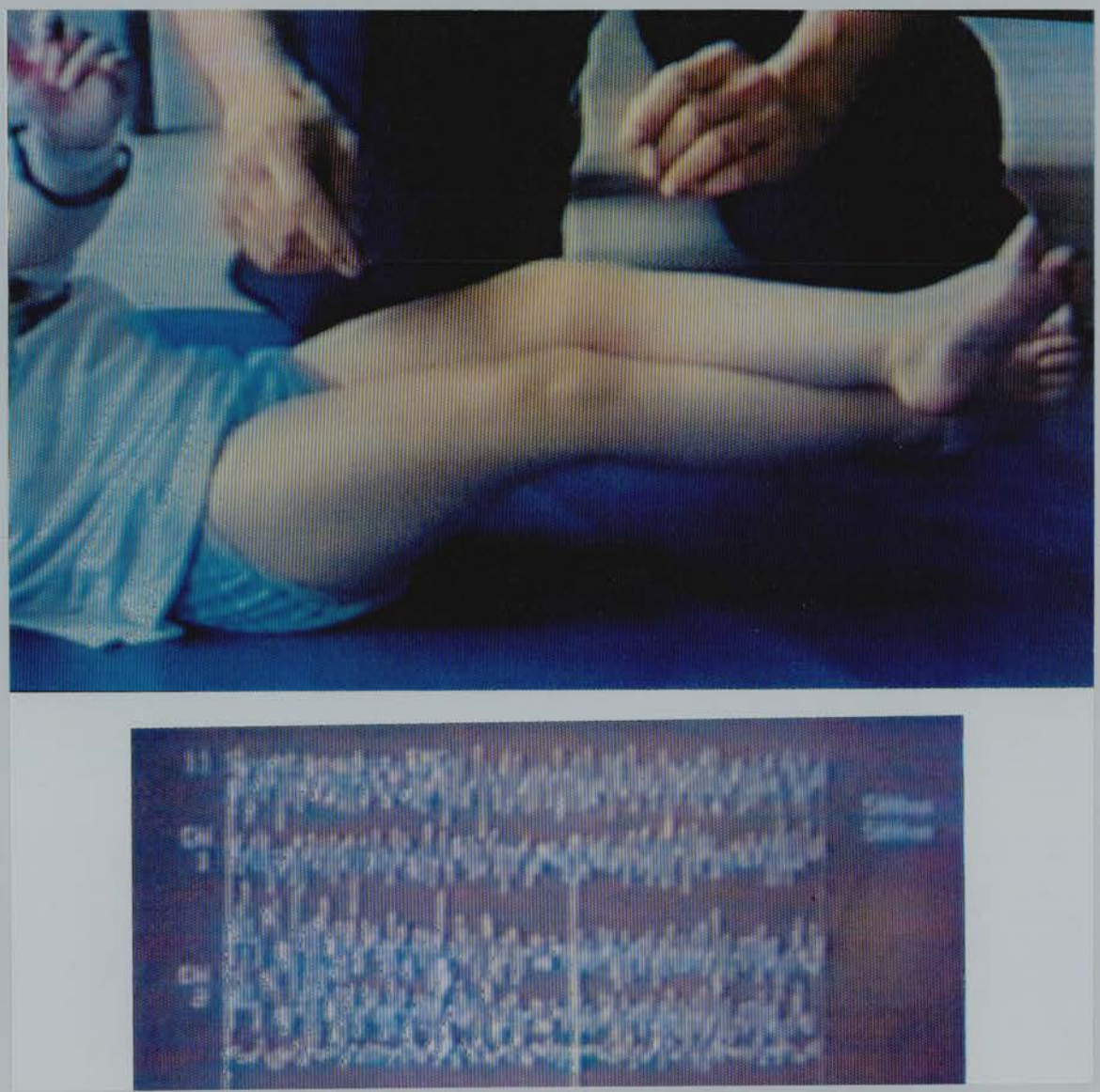

Figure 10.3.1 Effect of supine position on adductor and extensor posture in diplegia.
Three-year-old boy with diplegia of prematurity and shunted hydrocephalus secondary to intraventricular haemorrhage.At the time of this examination he had just developed a reciprocal crawling pattern and could sit unsupported, but was unable to stand self-supported or pull to stand. His preference was for 'W' kneeling which accentuated the tendency to internally rotate the legs at the hip. Hand function appeared well preserved as was speech and he was seizure-free.
Top: The legs are markedly adducted and internally rotated at the hip, extended at the knee and in equinovarus at the ankle. Note the spontaneously extensor toe. The leg tone fluctuates markedly with excitement and anticipation and a variety of other non-specific stimuli. Passive flexion at the knees is impossible. The knee jerk is not brisk.
Bottom: EMG recordings from the thigh muscles indicate continuous, high amplitude discharges 'at rest'.

Figure 10.3.2 Effect of vertical suspension on leg adductor and extensor posture in diplegia. Top: The arms are flexed, the legs are typically scissored, and extended at the knees, with equinovarus at the ankle.Bottom: Attempted passive abduction of the legs results in the whole body following the line of pull 'of a piece', with no obvious abduction possible.

Figure 10.3.3 Effect of updside down position on adductor and extensor posture in diplegia.
Top: The legs are flexed and abducted at the hips, flexed at the knees and the equinovarus of the feet is less marked. The arms are more extended.
Bottom: Sagital plane view shows the hip, knee and ankle flexion.

Figure 10.3.4
Effect of sleep in the supine position on hip adductors and internal rotators in diplegia.
Top: With sleep, the legs externally rotate and abduct at the hip. The big toes are no longer extended and passive motion can be performed with ease as the legs are limp.
Bottom: The EMG recordings from the thigh muscles are now silent.The effect of sleep is to abolish the tonic labyrinthine input and the dystonia.

As the figures illustrate, tonic adduction and internal rotation of the legs at the hips, extension at the knees, equinovarus at the ankles and spontaneous extensor toes are accentuated in the supine and upright positions, and relieved by

i. inversion (being held upside-down)

ii. sleep.

When awake, these muscles are continuously active (fig. 10.3.1), and EMG discharges surge according to the level of non-specific arousal, such as when being spoken to, speaking, using the hands, as well as with alterations of the child's position in space. It is impossible to obtain a passive joint range in the awake state. When asleep (fig. 10.3.4), the surface EMG recordings of the thigh muscles falls silent and the legs abduct, assume an externally rotated posture and feel floppy. Passive motion can be performed at any joint. Sleep appears to have completely abolished the tonic labyrinthine contribution to posture.

Clearly, the postures and extensor toes vary according to position in space, mental state and level of arousal.

10.4 Tonic labyrinthine reflexes, dystonia and sleep.

The awake postures in figures 10.3.1-3 above are evidently abolished by sleep (fig. 10.3.4) and exacerbated by the supine and erect positions. The inhibitory effect of REM sleep on most dystonic states has been extensively studied by Fish and colleagues (1991) as well as in the dyskinetic movements of Parkinson's disease, Gilles de la Tourette syndrome and Huntington's disease (Fish *et al*, 1991).

Sleep and arousal have an obvious and marked effect on motor behaviour. The H-reflex, which has been the brief object of interest previous sections (section 7) depends on the level of arousal and increases during alertness (Soriano *et al* , 1995; Schieppati, 1987), decreases during sleep and can be abolished during rapid eye movement sleep (Pivik and Mercer; Hodes and Dement, 1964). Clearly Ia afferent transmission and supraspinal motor control are altered by sleep.

What is interesting in this demonstration, is the abolition of the tonic labyrinthine reflexes by sleep.

10.5 Spontaneous extensor toe in infancy and cerebral palsy: an index of dystonia?

Another impressive feature of the diplegic child in addition to the "scissoring" demonstrated above, is the constancy of the spontaneously extended great toes, enlarged in figure 10.5.1. But is this sign the same as the Babinski sign or is it different? Figure 10.5.2 shows spontaneous extensor toes in a normal infant in the absence of a nociceptive stimulus, suggesting that it may reflect a physiological dystonia of supraspinal origin. According to Dittunno and Bell (1996) writing about the 100th anniversary of the eponymous sign:

"Today it is well accepted that the Babinski sign is indicative of dysfunction of fibres within the pyramidal tract (van Gjin, 1996)The fibres that control the foot and toes are the most likely to be dysfunctional if the Babinski sign is present, and over 90% of patients will have some degree of motor deficit in the foot (van Gjin, 1978)."

Figure 10.5.1 The spontaneous extensor toes in diplegia: a sign of spasticity or dystonia?
Same patient as for fig. 10.3.1. The toes relaxed when the child fell asleep, fig. 10.3.4. The extension of the great toe is not provoked by a nociceptive stimulus, but seems to be under supraspinal influence.

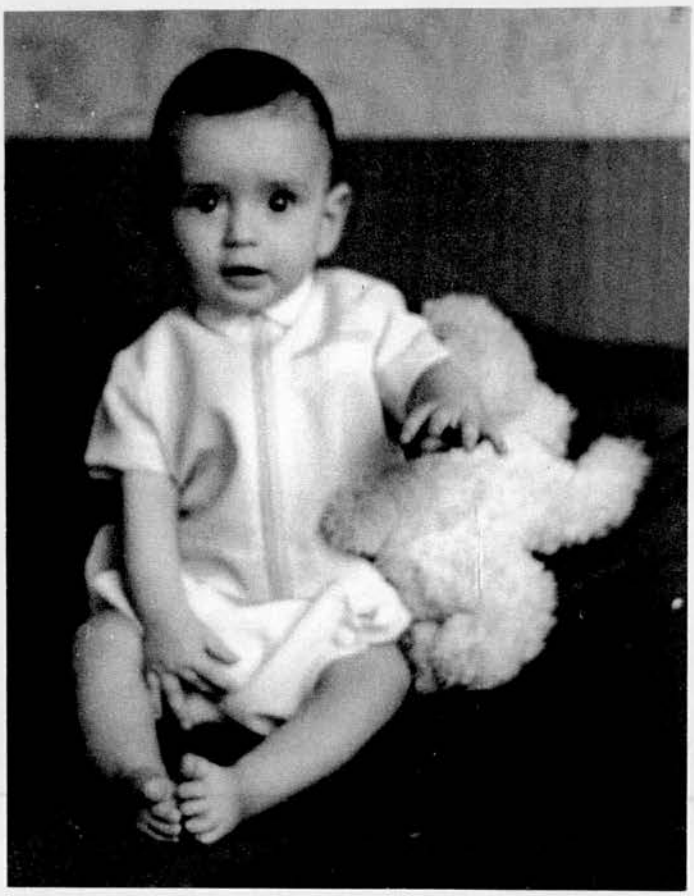

Figure 10.5.2 Spontaneous extensor toes in normal infant: ?physiological dystonia.
This normal infant is sitting unsupported and developed normally: there is spontaneous extension of the great toes bilaterally in the absence of a nociceptive stimulus.

In babies and infants, the presence of the clinical Babinski sign has been accepted as normal, and the extent to which this obtains has recently been reviewed (Bodensteiner, 1992), with a wide disparity among studies in the method of its elicitation:

"The position of the subject is also of importance in the evaluation of the plantar response, the patient should be in the supine position with the leg extended and the ankle at 90° of flexion. If the patient is sitting, the knee must be extended and the ankle flexed in order to best elicit the response. If the sitting patient is allowed to bend his or her knee, the Babinski sign will be lost in 50% of the patients who would otherwise have a positive Babinski sign. In studies of the plantar response in normal newborn infants, the position of the subject is clearly specified in only two (Hogan and Milligan, 1971; Rich, Marshall and Volpe, 1973). Both of these studies found that more than 90% of normal infants had flexor plantar responses. However a submaximal stimulus (mildly nociceptive or even non-nociceptive) was used to elicit the response in these studies. "

Bodensteiner, 1992.

Referring to the work of Landau (1987), Bodensteiner adds:

"Although lesions that produce Babinski signs involve the upper motor neurone pathway, including , but not limited to the pyramidal tract, the Babinski sign may be an indication of suppression (physiologic) of pyramidal function. The Babinski sign is also to some extent "state dependent" in that it can be seen with deep sleep, drug intoxication, postictal states, metabolic encephalopathies, posttraumatic encephalopathies (acute and chronic), and chorea and/or athetosis (may produce an apparent response not due to reflex response but due to posturing) "

Bodensteiner, 1992.

The clinical findings demonstrated in 10.3. 1-4, above, have been reproduced in the Movement Disorders Clinic at the Royal Hospital for Sick Children in Edinburgh and Movement Therapy Clinic at Guy's Hospital in many children each week, and are therefore not a negligible manifestation of diplegia. The histories of tone reduction or even floppiness in sleep are common accompaniments to these findings. The effect of sleep appears to be important since in the child described in 10.3, above, sleep *abolished* the spontaneous extensor toes whereas in the accounts by Landau (1987) and Bodensteiner (1992), the Babinski sign emerges in deep sleep. *If the spontaneous extensor toe in diplegia were a manifestation of the Babinski sign, why should it disappear in sleep instead of becoming more pronounced?* The point has been laboured since it is common to refer to 'spastic diplegia' as being the commonest manifestation of the cerebral palsies of prematurity, (see section 1, above) and since powerful 'antispastic' therapies are frequently advocated for these children, Peacock and Staudt (1990), and at increasingly earlier ages, for example, Park *et al* (1992).

It seems important to define the relative contributions of genuinely spastic

phenomena, that is to say pathological states which depend on Ia afferent input, and non-spastic components of the motor disorder which rely on other mechanisms for their maintenance, such as the plastic muscle change described in section 4, tonic labyrinthine and neck reflexes and Føg's posturing described above.

As discussed in the opening sections, the term "spasticity" is often used synonymously with that of "cerebral palsy", to the extent that in the absence of frank athetosis, dyskinesia or ataxia, all children with cerebral palsy are deemed to be 'spastic' unless proven otherwise.

An example of the confusion arises even in texts purporting to advise clinicians on a means of assessing potential candidates for surgery:

"Spasticity has been defined as a motor disorder characterised by a velocity-dependent increase in the resistance to passive stretch associated with hyperactive tendon reflexes (Lance, 1980). It is one feature of the upper motorneurone lesion that may occur at any level of the brain or spinal cord. Spasticity is characterised by: 1). Hypertonia of the clasp-knife type in which resistance to passive stretch gives way with continued force, as opposed to rigidity where resistance is felt throughout the range of motion; and 2). exaggerated tendon reflexes with or without clonus. Associated features of the upper motorneurone lesion may include: 1). a loss of isolated muscle control and fine voluntary movements; 2). weakness; 3). lack of normal postural reactions; 4). presence of abnormal associated movements; 5). depression of superficial reflexes; and 6). presence of specific abnormal reflexes {for example, Babinski, Oppenheim(Samilson, 1975)} "

Oppenheim et al 1992, p 276

These, definitions have been discussed in the preceding sections, but it should be noted that that Lance's operational definition of spasticity is employed. The difficulties arise not with the operational definition but with the syndromic definition of spasticity, which, as discussed in section 1.3.3: 'Classification of cerebral palsy', includes a number of other motor disturbances. Before returning to Oppenheim's text, it is worth reiterating the definition of spasticity offered by Crothers and Paine (1959) as a syndrome comprising:

1 Muscular hypertonus of the clasp-knife type
2. Hyperreflexia, which is reproducible on repeated elicitation
3. Positive Babinski and Hoffman reflexes
4. Diminished superficial reflexes (abdominal and cremasteric)
5. Release of postural and labyrinthine reflexes
6 Spread and overflow of associated movements
7. Loss of voluntary control of fine finger movements
8. Frequently clonus of ankle or other joints (although not universal or essential)
9. Tendency to muscular contracture in characteristic postures.

after Crothers and Paine 1959

The difficulty has become acute because of the availability of invasive and expensive treatments, which appear to require precise case-definition for selection. But to what extent

are practitioners of the new therapies able to clarify (or validate) their selection criteria?

"No surgical procedure can succeed unless it is directed towards specific goals and limited to those patients who have the actual potential to reach those goals. The goal of SPR (selective posterior rhizotomy) is to reduce spasticity. Clearly then, the best candidates for SPR are purely spastic children who had a low birthweight and were born prematurely. Such children are usually identified by the age of one year. They normally display an initial hypotonic stage that progresses to frank spasticity, with persistence of primitive reflexes and other features of the upper motorneurone lesion as enumerated above. Full-term children are more likely to have dystonia or athetosis along with the spasticity and are less responsive to SPR. The ideal patient is an intelligent, motivated child with spastic diplegia and no severe contractures, who is already walking independently and is attempting to improve his/her gait pattern and endurance. SPR does not create the potential for ambulation, which is the primary goal of many families. However , SPR can improve the gait of the child who is walking or developing that skill. Bleck's criteria for ambulation prognosis are useful as guidelines. "

"Other candidates for SPR are severely involved quadriplegic patients whose spasticity interferes with comfort, sitting, dressing, perineal care, or various classroom activities. Improvement in upper extremity function and speech are not primary goals of the procedure but may accompany an overall reduction in tone. Moderately-affected children with spastic cerebral palsy should be very carefully evaluated because many factors other than spasticity interfere with function"

Oppenheim et al 1992, p 276

Several issues are raised in this brief statement on the selection of suitable candidates for selective posterior rhizotomy which give cause for concern, not least the emphasis on selecting "purely spastic children", the most able-bodied and the most severely impaired children, and the apparent caution against the selection of the moderately impaired children "because many factors other than spasticity interfere with function": does this imply that severely affected children do not have other factors that interfere with function? It is also notable that in the above statement, 'frank spasticity" and "the persistence of primitive reflexes and other features of the the upper motorneurone" form part of the inclusion criteria for this antispastic therapy. However, on the same page, the author describes the criteria for rejecting children:

"Because cerebral palsy is a multifaceted disorder, assessment for patient selection should emphasise the identification of features of cerebral palsy that will persist following surgical reduction of spasticity. First the evaluation team determines that spasticity is present and interfering with function. Other forms of abnormal tone and movement should be identified so that patients with rigidity, dystonia, athetosis, ataxia, and truncal hypotonia can be eliminated as candidates. Rigidity is characterised by a "lead pipe' type of resistance to passive motion. Dystonia involves fluctuating tone, often associated with primitive reflexes. Athetosis is punctuated by involuntary movement around the a joint axis, orofacial movement and finger fanning. Ataxia is recognised by by the findings of disequilibrium, intention tremors and and past-pointing, In those with mixed lesions, the results will be mixed. Patients with pure spasticity are the best candidates."

Oppenheim et al, 1992, p276

Quite clearly, the statement that "dystonia involves fluctuating tone, often associated with primitive reflexes", links the relationship between dystonia and persistent primitive reflexes, whereas in the statement on positive criteria for selection, persistent primitive reflexes were included in the definition of the spastic syndrome.

This lack of clarity is unfortunately both a product of the history of neurology and the inherent difficulty in making water-tight clinical definitions. Nor is the reliance on the absence of conspicuous basal ganglia lesions on neuroimaging, as indicated by Park and Owen (1992) and Rab (1992) a guarantee against the presence of clinical extrapyramidal involvement or dystonia, since, as has been made clear by Denny-Brown (1980), damage to the perirolandic regions produces extrapyramidal signs.

10.6. Associated movements and Føg's posturing.

In the introduction to the report in which the tonic neck and labyrinthine reactions are described, Walshe (1923), describes the phenomenon of associated movements:

"Neurologists are learning from Sherrington to regard muscle tone as the basis of posture, and decerebrate rigidity as a form of reflex standing. In the earlier analysis of spasticity, as it occurs in hemiplegia and in the extended form of spastic paraplegia, reasons were given for regarding it as physiologically identical with experimentally produced decerebrate rigidity. It was pointed out that all voluntary purposive movements are accompanied by an appropriate postural adjustment of the rest of the skeletal musculature, and that in forceful movements this adjustment or adaptation is necessarily bilateral and widespread. Although carried out under voluntary control, postural adaptation is a function of reflex mechanisms situated in the brain-stem, which are not put out of action by the lesion which produces hemiplegia and abolishes voluntary control of the musculature on the affected side of the body. In these circumstances, we should still expect postural reactions to occur when forceful voluntary activities are carried out by the musculature of the sound half of the body. Now, however, deprived of cortical control, they would occur in exaggerated intensity and deprived of that fineness of adaptation which that control ensures. It is was concluded that the "associated movements" of hemiplegia are phenomena of this order, appearing in the muscles of the affected side on certain voluntary movements of the normal limbs, or on such semi-voluntary movements as yawning. In other words associated movements, or, as we shall call them, "associated reactions" are released postural reactions in muscles deprived of voluntary control."

FMR Walshe, 1923.

Walshe (1923) describes a variety of postures adopted by the affected limb during voluntary forceful isometric tasks of the nonparetic limb. In section 3 on the mechanisms of equinus above, an example of the running posture in two children with hemiplegia is given, illustrating the emergence of a normal functional posture for running in one of the cases (figure 3.4.6d) when in the same case the walking posture was abnormal (figure 3.4.6c). In the other case, a 12 year old boy with a left hemiparesis, the running posture of the hands looks awkward, with the fingers spread, extended at the metacarpophalangeal joints and the

right wrist supinated (fig.3.4.6b). Nevertheless, the point must be made that in addition to the release of unusual or unwanted postures when changing from one physiological motor state to another (walking to running), a normal motor pattern may reemerge because, as was suggested in section 3, the running posture resembles an alternating physiological hemiposture (Lin and Brown, 1992).

Elin and Møgens Føg took up the study of associated postures and published their results on hundreds of healthy children and children with mental retardation in 1963. The point of interest being the persistence of associated movements in children without conspicuous evidence of a motor disorder as well as the exaggeration of such associated movements in the presence of established brain damage.

"In the mature and uninjured brain no associated movements occur when daily life activities or well-trained motor performances are executed. If, however, the brain is injured- especially if the damage has taken place in early life- or if an adult has to learn an unaccustomed and complicated hand performance, associated movements can be observed."

Elin and Møgens Føg, 1963

The tests which Føg and Føg devised were extremely simple:

"We have chosen two tests, one bilateral, feet to hands: and one crossed, from one hand to the other.
Feet-hands test: When a child inverts his feet, he will at the same time present associated movements of his hands, in most cases a supination, sometimes a pronation or extension.
Hand-Hand test: When a child exerts a certain degree of pressure with the thumb and first finger, a similar movement is observed in the opposite hand. Sometimes the reverse action takes place, the fingers being extended. We have described identical associated movements as a homologous reaction, and simultaneous extension as a heterologous reaction. Three degrees of pressure were tested using a spring clothes peg and two spring (bulldog) paper clips as tools"

Elin and Møgens Føg, 1963

The children comprised 265 healthy controls aged 2-16 years and 184 mentally retarded children aged 8-16 years with I.Qs ranging from 60-95, 117 who had "no history indicative of cerebral lesion" and 67 in whom there was a history of birth complications or post-natal injuries.palsy.

The results remain startling today. For the feet-to-hand test, associated supination of the wrist with foot inversion persisted in 75% of healthy 8-10 year-olds, and some form of associated posture in almost a third of 14-16 year olds. For the hand-to-hand tests, homologous movements were present in 75% of 5-7 year olds,over half of the 8-13 year olds and up to a third of the 14-16 year olds, especially when using the stiffer springs. For the

feet-hands test, the mentally retarded children showed a persistence of some form of associated movement in all of the 16 year-olds with a history of motor damage and 75% of those without such a history, compared to just over a quarter of healthy 16 year olds. The disparity between groups being more marked in the hand-hand tests

Føg and Føg concluded that associated movements are normally progressively suppressed during the course of childhood, and that "they are more so in normal than in mentally retarded children" and that the persistence of such movements could indicate minimal brain damage or a "poor development of discriminatory, selective motor activity.".

10.7 An example of mirror movements of first dorsal interosseus muscles in congenital hemiplegia during graded isometric contractions.

One of the crucial elements of the observations by Walshe (1923) and Føg and Føg (1963), is that the associated movements involuntarily accompany intended movement.

This is shown in figure 10.6.1 in a boy with a left hemisyndrome: top, walking normally with a heel-strike on the left; middle, voluntarily toe walking, which produces pronation at the wrist, flexion at the metacarpophalangeal joints and extension at the interphalangeal joints ie 'hand and finger equinus"; bottom, voluntarily heel-heel walking during which the left wrist is markedly extended as are the fingers, although this affects the right wrist to a lesser degree.

Associated or mirror movements can be documented in a variety of ways (Woods and Teuber, 1978; Farmer et al 1990, 1991; Carr et al, 1992, 1993) and are illustrated in figure 10.6.3 showing the isometric force exerted by the left (nonparetic) first dorsal interosseous (FDI) muscle of the left hand with simultaneous recordings of the surface EMG from the FDI muscles of both hands. Clinically, the right arm was seen to participate in bimanual tasks such as clapping and holding the handlebars of a tricycle in the second year, but always requiring initiation of such tasks with the left hand first.At the time of investigation, there was obvious right-sided hemiatrophy. Attempts at using the right hemiparetic hand alone produced only minimal shrugging movements at the right shoulder and elbow.and gross finger extension with the long extensor muscles of the forearm. Use of both hands produced marked mirror movements with the ability to form a pincer grip, repetitive thumb and index opposition and crude sequential opposition of the thumb with the other digits on the hemiparetic side (Denckla, 1973).

These movements were performed at the same frequency as on the left but with a

smaller amplitude. The mirror movements correspond to a Woods and Teuber (1978) grade 3 on a scale of 0-4. In addition to the mirroring of fine distal hand and finger tasks when movements were initiated with the left hand, rapid alternating movements such as flexion and extension at the metacarpophalangeal and wrist joints, pronation-supination at the wrist, flexion-extension at the elbow and abduction-adduction at the shoulder were all possible at the same frequencies as on the nonparetic side, though the amplitudes of movement were reduced on the hemiparetic side.

Although some proximal movement at the shoulder and elbow joints could be generated alone, he was unable to perform unilateral abduction of the right index which became possible only with bilateral index finger abduction. Figure 10.6.2 shows the experimental arrangements for recording the isometric force and surface EMG from the FDI (in this instance recording from the right hand). In figure 10.6.3 the subject is instructed to maintain the lowest steady abduction force output with the nonparetic index, using the dial and LED lights to guide him (fig. 10.6.2). He was then asked to gradually increase the steady (isometric) force every 50 seconds or so, to produce a step-wise pattern of increasing force on the computer monitor.

It can be seen that at very low isometric forces of about 0.25N , the motor units recruited are "small" in amplitude and that each isometric increment is met by an increase in the rate of firing and size of the motor units participating in the task, as recorded by the surface EMG electrodes. The ability to increase abduction force level in the normal hand is underpinned by a gradual increase in firing frequency of the active motor units and recruitment of motor units of increasing apparent size. The slight variation in the abduction force output trace is due to fluctuations in the sustained motor unit firing frequency.

The overall effect is that of a "crescendo" pattern of motor unit firing from the nonparetic FDI, the so-called *Henneman Size Principle* (see Henneman 1974, Jones and Round, 1990) of motor unit firing frequency and recruitment. The raw EMG recorded from the hemiparetic FDI muscle shows little or no activity for the first 50 seconds, but greater activity as the nonparetic index abducts at higher force levels: in fact, from 50 to 225s of the recording, the hemiparetic FDI muscle performs an associated recruitment of motor units.

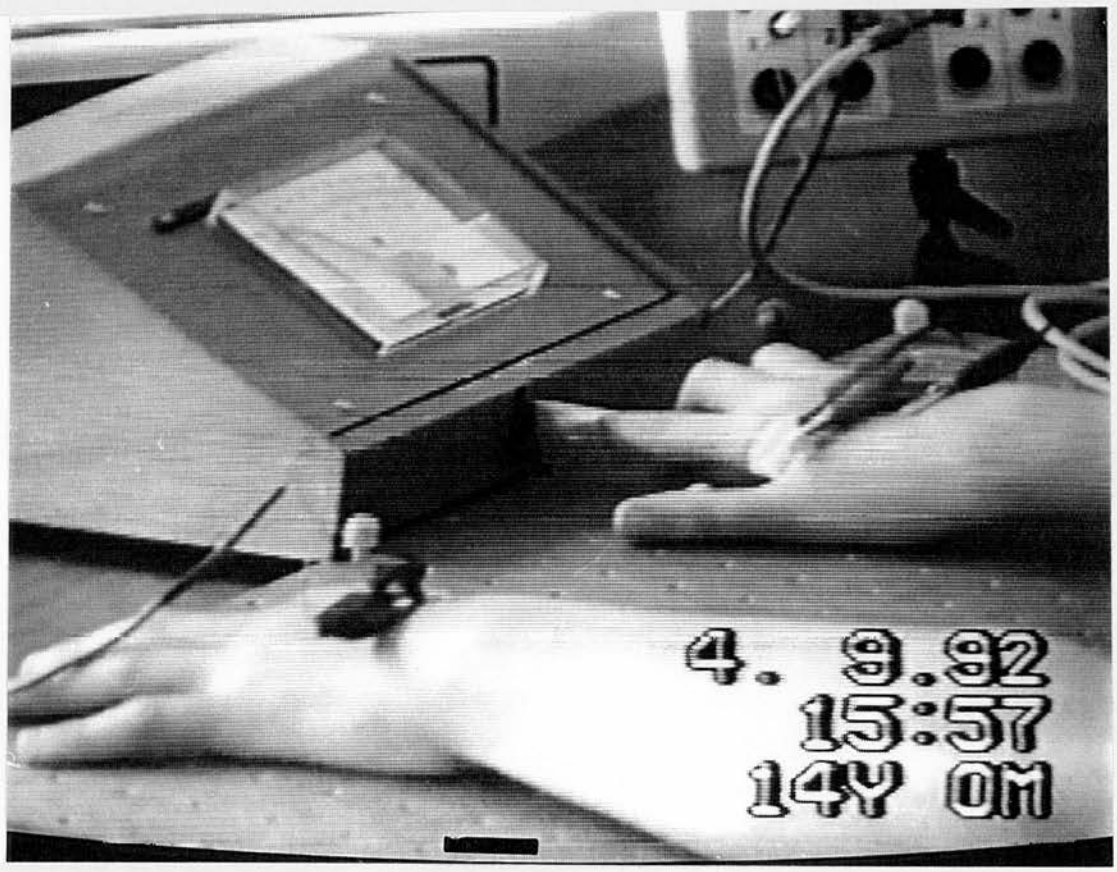

Figure 10.6.2
Arrangements for recording of isometric surface EMG and abduction force from the first dorsal interosseus (FDI) muscle of the index finger of the right (hemiparetic) hand.The subject is informed of the strength of the isometric force by coloured LCD lights and the force is also displayed on a dial. The force plate is bidirectional so that the isometric FDI muscle force of either hand can be measured.

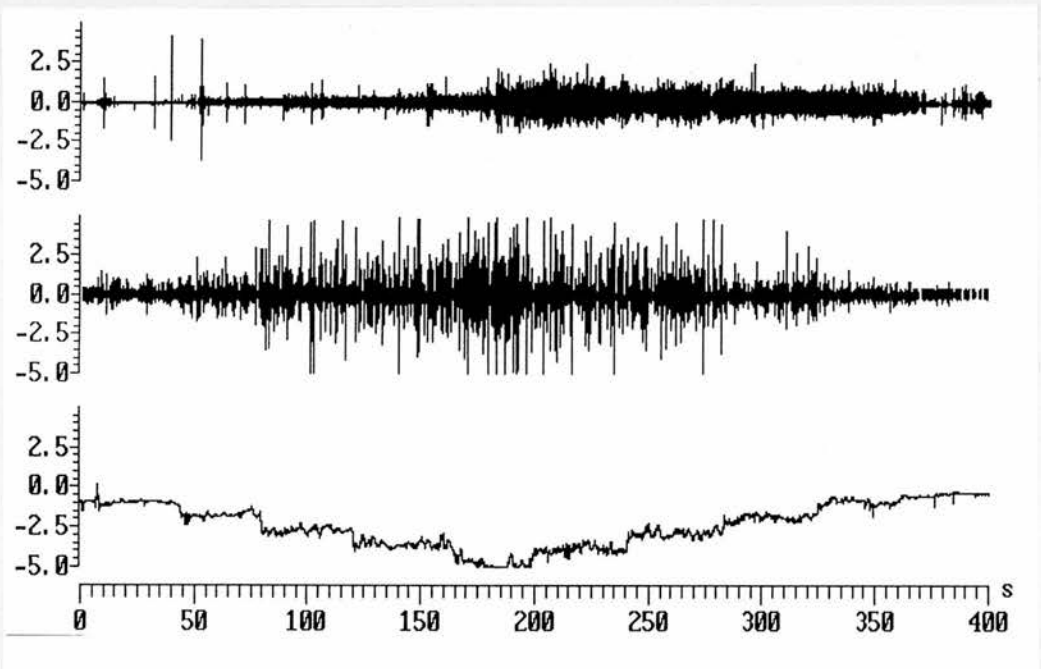

Figure 10.6.3. Index abduction task:
Voluntary isometric finger abduction with increasing force output from nonparetic first dorsal interosseus (FDI). Top trace: Hemiparetic surface EMG from right FDI muscle.
Middle trace: Nonparetic surface EMG from left FDI muscle. Bottom trace: Isometric force output from nonparetic (left) FDI muscle.
i) The force trace shows five consecutive steady increases in isometric force. Small forces of 0.5 N can be maintained, building up to 2N. **ii)** Each steady isometric increment is accompanied by an increased rate of firing and recruitment of apparently larger motor units to produce a crescendo pattern of motor unit firing. **iii)** Note that initially, there is little or no EMG recorded from the hemiparetic FDI, though this builds up as the nonparetic FDI exerts greater force. **iv)** The subject felt no particular effort in sustaining a graded increase in isometric force output from the nonparetic FDI, in marked contrast to the extreme effort required when a similar task was attempted with the hemiparetic FDI. Horizontal scale=seconds.

When the hemiparetic index is attached to the force plate and the subject invited to abduct this finger in isolation, no output is possible as witnessed by the absence of motor unit spike discharges from the hemiparetic FDI muscle until the nonparetic FDI is allowed to participate in the task, implying that control of the right FDI depends on prior activation of the nonparetic FDI muscle.

Many of these common observations have been reviewed by Brown and colleagues (1997) who have included observations on the postures of the foetus, newborn (preterm and term) infant and child in health and disease to emphasise the developmental aspect of posture which has been extensively discussed in section 3 on the mechanisms of equinus above. The extent to which posturing of the hands during an equinovarus gait might change after peripheral intervention to correct the foot has not been fully addressed to date.

The main reason for including this brief mention of tonic labyrinthine, Føg and associated posturing and mirror movements is to highlight additional, 'centrally driven', 'non-spastic' motor strategies.

Figure 10.6.1 Føg posturing during walking.
Top: normal gait, hands normal.
middle voluntary equinus gait, hands in equinus.
bottom: voluntary heel-heel gait, hands follow suit, especially on the left.

10.8 Summary.

i. Tonic labyrinthine influence on posture in diplegia of prematurity has been demonstrated. The supine and vertical position accentuate hip adduction and internal rotation, extension at the knee and equinovarus of the feet, together with spontaneous great toe extension, all of which are accompanied by continuous, high amplitude electromyographic (EMG) activity in the legs

ii. A reverse posture obtains when the child is held upside down: namely legs abducted and flexed at the hip, knee and ankle.

iii The tonic labyrinthine reflexes are abolished by sleep, as is the EMG activity, which is electrically silent. During sleep, the legs are abducted and externally rotated at the hip, and the great toes are flexed.

iv The Babinski response is enhanced by sleep.

v The dystonic extensor toe is relieved by sleep: it is not the same as a babinski responsewhich can only be elicited by a nociceptive stimulus to the sole of the foot.

vi Dystonic postures may be part of normal and abnormal development and dystonic postures are invariably abolished by sleep.

vii Spasticity itself produces no postures.

vii Associated postures accompany movements and are usually more pronounced during motor development. They may reappear in the mature motor system during the performance of unaccustomed tasks; they may persist or be released in the presence of brain injury: in all cases (apart from the primitive reflexes emanating from neck or limb sensory afferents) they are of supraspinal origin and are distinct from spasticity.

viii. Associated movements and postures of the hands during voluntary toe-toe or heel-heel walking are demonstrated as examples of developmental immaturity which persist in the presence of neurological impairment.

ix. Associated and mirror movements accompany voluntary tasks and are different from dystonia or spasticity

x. The implications for classification and selection for treatment (s), with particular emphasis on the selection for antispastic treatments, is discussed.

11. End note.

Different aspects of the motor manifestations in children with cerebral palsy have been reviewed, examined and measured noninvasively.

The epidemiology, case-definition and known natural history in cerebral palsy have been reviewed along with the rationale and published results of some of the more invasive treatments currently available.

The mechanisms of hypertonus have been examined in detail and expanded to encompass much more than 'spasticity', necessitating a detailed examination of the physiological basis of: intramuscular 'plastic' transformation; abnormal walking patterns such as equinus; stretch reflex excitability thresholds and gain; reflex muscular twitches and the optimal neuromuscular angle; muscle-twitch phenotype and malleability in response to internal and external mechanical stimuli and the variable expression of a reentrant reflex phenomenon such as 'clonus'. The relationship between central and peripheral structures and functions is emphasised again and again, with the bulk of this emphasis on the muscle itself.

Measurements of the speed of alternating movements have been made in an attempt to demonstrate the physical boundaries and principles governing such tasks as well as the neuromuscular adaptations required to accomplish them. Developmental aspects are touched upon linking the relationship between bone growth, increasing limb inertia and hence demands on the muscular system. Once again, muscle growth in length and cross-sectional area as well as muscle-twitch phenotype appear to contribute to the development of an increased force and speed which in turn contributes to the maturation of motor dexterity along with selection of the appropriate functional neuromuscular joint angle.

If muscle length, in part, determines force output and reflex excitability, a clear link between posture and mobility can be established. The effect of normal and abnormal postures may be predicted according to the peripheral responses of the neuromuscular system. Accordingly, one aspect of the lack of motor dexterity in hemiparesis results from the neuromuscular disadvantage imposed on the motor system by an unfavourable posture; a disadvantage which may become permanent if the muscle develops a contracture. Unopposed gravity, disuse and immobility, by producing contractures impair function, which favours further contracture.The benefits of strengthening as opposed to weakening muscle

is strenuously advocated in the body of this text.

Postures such as the release or failure of inhibition of tonic neck and labyrinthine postures; dystonia; running postures; associated postures, Føg's posturing and mirror movements and all represent different facets of the ontogeny or arrest of motor development in cerebral palsy arranged in a hierarchy of apparent increasing complexity. Their inclusion as aspects of 'spasticity' results in an inevitable confusion when interpreting 'antispastic' treatments, since none of these entities would be influenced by dorsal root rhizotomy. On the other hand, all of these would be affected, one way or another, by botulinum toxin A, since the site of action is muscle itself.

In one main respect, these studies of different aspects of motor function and control emphasise the need to anticipate changes in motor function and to incorporate a number of strategies into any treatment regimen to maintain function or prevent the loss of residual function.The mere doling out of infrequent, one off 'treatments' may be administratively and financially attractive but is at best a delusion and at worst, a complete misunderstanding of the behaviour of the motor system.

It is anticipated that a greater emphasis on exercise and fitness-training with judiciously applied orthotic, medical and surgical treatments aimed at restoring positions of function offer the best hope of maintaining and promoting functional independence in children with cerebral palsy. Treatments should be based on a proper physiological understanding of maladaptive postures and movements, of central and peripheral interactions between the elements of the motor system and of the insidious consequences of immobility and disuse.

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Original papers

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Heterogeneity of neurological syndromes in survivors of grade 3 and 4 periventricular haemorrhage

Jean-Pierre Lin, Winnie Goh, J. Keith Brown, A. James W. Steers

Department of Paediatric Neurology, Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF, UK

Received: 15 October 1992

Abstract. To evaluate the topographical neurological distribution, patterns of abnormal tone and related functional neuromotor impairment after grade 3 and grade 4 intraventricular/periventricular haemorrhage (IPVH), 33 children with previous grade 3 or 4 IPVH of mean gestational age 30.9 weeks (range 25–40 weeks) and mean birth weight 1743 g (range 866–3600 g) were examined neurologically at 4.7 years (range 0.75–10.8 years). Neurological signs were absent in 10/33 cases which were equally distributed between the grade 3 and grade 4 IPVH groups. The largest single topographical neurological distribution was hemiparesis in 8/23, followed jointly by diplegia (cerebral paraplegia) in 6/23 and triplegia in 6/23 cases and finally quadriplegia in 3/23 cases. Grade 4 IPVH tended to result in asymmetrical syndromes, accounting for 7/8 cases of hemiparesis and 5/6 cases of triplegia, whereas all 3/3 cases of quadriplegia followed grade 3 IPVH. The 6/23 cases of diplegia were shared between the grade 3 and grade 4 IPVH groups. Tone was normal in 7/8 of the hemiparetic subjects. Dystonia was the commonest tone abnormality, affecting 8/23 children with neurological disturbance, followed by ataxia/hypotonia in 4/23 and mixed dystonia/hypotonia in 3/23. Only 1/23 cases had signs of spasticity. Spasticity is rare following severe IPVH. Diplegic children had a better functional neuromotor grade than hemiparetic children, who in turn did better than triplegic children. Ataxia hypotonia resulted in better functional outcome than dystronia, which in turn was more favourable than mixed tone patterns. Cranial imaging by ultrasound (US) or computed tomographic (CT) scanning proved an unreliable prognostic indicator except in the case of hemiparesis, for which US scans correctly predicted the affected side in 5/7 cases. The neurological syndromes following severe IPVH differ from the classical encephalopathy of prematurity, and this should lead to a re-appraisal of the trends in the prevalence of cerebral palsy. Caution should be exercised in the interpretation of cranial imaging with regard to pessimistic prognoses in the presence of changes or undue optimism in their absence.

Key words: Intraventricular/periventricular haemorrhage – Topographical neurological distribution – Tone – Functional outcome

Correspondence to: J.-P. Lin

Introduction

Cerebral palsy (CP) is defined as the motor manifestation of a non-progressive injury to the developing brain. It excludes degenerative diseases and conditions affecting the spinal cord, peripheral nerves and muscles. The cause of the majority of cases of CP is unknown, and its overall prevalence remains unchanged at 2–2.5/1000 in industrialised countries despite marked changes in obstetric and neonatal care [17]. One characteristic CP syndrome which has declined in incidence is the dyskinetic CP associated with Rhesus incompatibility and kernicterus. This is due entirely to antenatal screening and the policy of protecting future pregnancies by offering at-risk mothers anti-D shortly after delivery. Since such a screening policy can help eradicate a whole class of CP conditions, the logical way forward for research into the prevention of CP is the definition of distinct neurological syndromes within the CP population to distinguish between CP of genetic, antenatal, perinatal or postnatal origin. Such syndromes would help focus programmes for prevention or intervention.

Despite an apparent common aetiology and pathology, the neurological syndromes following IPVH are very diverse and opinions remain divided regarding the exact mechanisms and timing of the haemorrhagic insult [16]. It follows that many other conditions with specific aetiologies may result in differing pathologies which produce varied neurological signs leading to several clinical syndromes.

Aims

We have previously described the neuromotor outcome of 33 children with severe IPVH in terms of their perinatal histories, neonatal course, pre-operative raised intracranial pressure, shunt complications and number of shunt procedures [11]. This article attempts to illustrate the heterogeneity of neurological manifestations following grade 3 and grade 4 IPVH and compare these findings with those of the encephalopathy of prematurity.

Method

All of 33 cases of grade 3 or 4 IPVH in children born between 1975 and 1988 available for follow-up in the Department of Paediatric Neurology of the Royal Hospital for Sick Children, Edinburgh, underwent a neurological examination. Cases with associated major congenital malformations, chromosomal defects or gross periventricular leucomalacia had been excluded on the grounds that the primary effects of IPVH would be obscured. The children were examined in terms of their functional neuromotor grade [11], topographical neurological distribution of signs and the patterns of abnormal muscle tone.

Definitions

Topographical neurological distribution. All cases were examined and allocated to one of four topographical types:

Normal:	No neurological signs in any limb
Hemiparesis:	Neurological signs confined to an arm and a leg
Diplegia:	Neurological signs confined to the legs only
Triplegia:	Neurological signs affecting one arm and both legs
Quadriplegia:	Neurological signs affecting all four limbs

Muscle tone. Definitions of muscle tone follow the classification of Brown and Minns [3] and are summarised as follows:

Phasic spasticity: Resistance to passive movement of a limb at a joint in response to *rapid stretch* of the muscle

Dystonia: Fluctuations in tone with position in space and body contacts characterised by *spontaneous extensor toe responses*, legs in extension, internally rotated and adducted at the hip, with or without scissoring

Ataxia/hypotonia: Reduced tone on *passive stretching*. Legs internally rotated and abducted at the hip in stance with a valgus tilt at the ankle joints during weight-bearing

Mixed tone: Any combination of phasic spasticity, dystonia or ataxia/hypotonia

Functional neuromotor grade. This has been defined in our previous report [11] as normal, mild, moderate, severe or profound functional impairment.

Encephalopathy of prematurity or Little's disease. Encephalopathy of prematurity, also called Little's disease, occurs in preterm children without an overt acute clinical neurological illness in early life such as asphyxia, fits or IVH. It was clearly described by Polani in 1958: "The post-prematurity spastic paraplegia type of cerebral palsy is a striking clinical entity, striking for the symmetry of the neurological signs, for their distribution, for the relatively good intelligence of the patients, and the comparative freedom from seizures; and all these features tend to militate against the idea of brain injury being responsible." [14]. The features of the early disturbances in tone attributable to uncomplicated prematurity or low birth weight have been previously described [1, 2] as a true rigid extension or dystonia of trunk and legs, accompanied by spontaneous extension of the big toes, motor delay of the lower limbs, with or without brisk reflexes and ankle clonus. If present beyond the 1st year of life, this may either resolve completely, persist as dystonia or merge into lower limb spasticity.

Ultrasound brain scans

These were performed at the time of the diagnosis by the neonatal team. Follow-up ultrasound (US) scans were performed serially as indicated by the clinical course during neonatal and subsequent neurological follow-up of ventricular size and management of hydrocephalus. Since all the children had ventricular dilatation, only the presence or absence of cysts is recorded.

CT brain scans

These were performed to document acute changes during the clinical management of the infants, but were not performed routinely at a predetermined age or after the first shunt. Three children had a CT scan without a prior US scan of the brain.

Results

Ten children were considered normal and the remaining 23/33 children exhibited a variety of motor impairments.

Topographical distribution of neurological signs and IPVH grade

The topographical neurological distributions (TND) and the underlying severity of IPVH are shown in Table 1. There were twice as many cases of grade 4 IPVH (parenchymal extension) as of grade 3. The 10 children with no neurological signs were equally represented in the grade 3 and the grade 4 IPVH groups. Of the 23/33 neurologically impaired children there were 8/23 with hemiparesis, 2 of whom were twin sisters with opposite sides affected; 6/23 had pure diplegia, 6/23 were triplegic and 3/23 quadriplegic. Asymmetrical neurological signs occurred more frequently in grade 4 IPVH, which accounted for 7/8 of the hemiparetic and 5/6 of the triplegic children. Children with symmetrical neurological signs, such as the 6 diplegic children, were proportionately distributed between the grade 3 and grade 4 IPVH groups but all 3 cases of quadriplegia arose from grade 3 IPVH.

Major pattern of disturbed tone and IPVH grade

Table 2 shows the major patterns of clinical tone in relation to IPVH grade. Tone was normal in 17/33 cases, which included 7 cases with hemiparesis, and 16/33 cases had abnormal tone. Only 1/16 children was thought to be spastic. Dystonia was the commonest pattern of disturbed tone, affecting half the cases with abnormal tone (8/16 cases), followed by ataxia/hypotonia in 4/16 and mixed patterns accounting for 3/16 cases.

Clinical assessment of tone and topographical neurological distribution

Comparison of TND and clinical tone (Table 3), 7/8 hemiparetic children had normal tone. All of the diplegic children had normal upper limb tone with signs of either dystonia (3/6), ataxia/hypotonia (2/6) or mixed tone disturbance (1/6) in the lower limbs. Triplegic children showed patterns of dystonia (4/6) or ataxia/hypotonia (1/6) only 1/6 having a mixed pattern. The only spastic child had a quadriplegic TND, the other two quadriplegics exhibiting dystonia and ataxia/hypotonia respectively.

Table 1. Topographical distribution of neurological signs according to IPVH grade ($n=33$). IPVH, Intraventricular/periventricular haemorrhage

Topographical neurological distribution	Grade 3	Grade 4	Total
Normal	4	6	10
Hemiparesis			
right	1	5	6
left	0	2	2
Diplegia	2	4	6
Triplegia			
right	1	3	4
left	0	2	2
Quadriplegia	3	0	3
Total	11	22	33

Table 2. Major pattern of disturbed tone according to IPVH grade ($n=33$)

Muscle tone	Grade 3	Grade 4	Total
Normal tone	5	12	17
Spastic	1	0	1
Dystonic	4	4	8
Ataxic/hypotonic	1	3	4
Mixed	0	3	3
Total	11	22	33

Functional neuromotor grade and topographical neurological distribution

Table 4 charts the functional neuromotor grade (FNG) in terms of the TND. All 8/8 of the hemiparetic children are classed as moderately impaired, whereas diplegic children have a broader range of FNGs: 2/6 with mild impairments of no functional significance, 3/6 a moderate and 1/6 a severe FNG. The triplegic children were more severely affected with 2/6 assessed as moderately and 4/6 as severely affected. All 3/3 of the quadriplegic children were classed as profoundly affected at the time of the study, but one of these has since developed wheelchair mobility which would put him among the severely rather than the profoundly impaired.

Functional neuromotor grade and pattern of clinical tone

The clinical assessment of tone against FNG is shown in Table 5, with 7/8 hemiparetic children having moderate functional impairment but normal tone. The only child with spasticity had a quadriplegic distribution of neurological signs and was profoundly impaired. Dystonic children showed functional impairments across the grading from mild to profound though 5/8 had moderate impairment of motor function. The ataxic/hypotonic group appeared to have less severe impairments than the dystonic group, while children with mixed patterns of tone had similar functional disturbances to the dystonic group.

Neurological signs and US scan or CT brain scan changes

All of the children had IPVH diagnosed on the basis of either a US scan or a CT brain scan. A total of 30/33 children underwent US scan brain imaging and 19/30 had CT brain scanning (Table 6). In only 9/23 of the children with subsequent neurological symptoms were cysts evi-

Table 3. Clinical assessment of tone and topographical neurological distribution ($n=23$)

Topographical neurological distribution	Dominant pattern of clinical tone					
	Normal	Spastic	Dystonic	Ataxic/ hypotonic	Mixed	Total
Hemiparesis						
right	5	0	0	0	1	6
left	2	0	0	0	0	2
Diplegia	0	0	3	2	1	6
Triplegia						
right	0	0	2	1	0	3
left	0	0	2	0	1	3
Quadriplegia	0	1	1	1	0	3
Total	7	1	8	4	3	23

dent on US scans performed in the neonatal period, which compared with 3/16 symptomatic children offered early CT imaging. Neonatal US scans in subsequently asymptomatic children were positive in 2/9 cases, compared with positive findings in 1/3 asymptomatic children offered CT scans. These findings are summarised in Table 7 for US scanning and Table 8 for CT scanning. Unilateral cysts were correctly identified by US scan in 5/7 children who subsequently developed a hemiparesis, but in only 2/6 hemiparetic children were lesions identifiable on early CT scans (Table 6.). Imaging by either US scan or CT proved poor in detecting future cases of diplegia. Half of the 6 triplegic children had bilateral cysts visible on US scans but in only 1/4 were lesions picked up on CT scanning. None of the 3 children who became quadriplegic had demonstrable lesions on US scan or CT scan other than the ventriculomegaly common to the group as a whole. Figure 1 shows the CT scan findings from five different children, three with hemiparesis, one with triplegia and one with quadriplegia, showing that brain morphology as seen on CT may not correspond with the neurological signs and clinical severity. Figure 2 demonstrates relatively good distal dexterity in a child with a right hemiparesis despite virtual destruction of the left hemisphere.

Table 4. Neuromotor functional grade and topographical neurological distribution ($n=33$)

Topographical neurological distribution	Functional neuromotor grade				
	Mild	Moderate	Severe	Profound	Total
Hemiparesis					
right	0	6	0	0	6
left	0	2	0	0	2
Diplegia	2	3	1	0	6
Triplegia					
right	0	1	3	0	4
left	0	1	1	0	2
Quadriplegia	0	0	0	3	3
Total	2	13	5	3	23

Concordance of initial US scan and subsequent CT brain imaging

Of the 19/33 children who underwent CT brain scans, 16/19 had previously undergone US scanning of the brain. The concordance between these two imaging techniques is shown in Table 9, irrespective of whether the scans were proved correct by subsequent neurological

Table 5. Functional neuromotor grade and the pattern clinical tone ($n=23$)

Functional neuromotor grade	Normal	Spastic	Dystonic	Ataxic/ hypotonic	Mixed	Total
Mild	0	0	1	1	0	2
Moderate	7	0	5	2	2	16
Severe	0	0	1	1	0	2
Profound	0	1	1	0	1	3
Total	7	1	8	4	3	23

Table 6. Topographical distribution of neurological signs and changes on US or CT brain scanning ($n=33$). US, Ultrasound; CT, computed tomography

Topographical neurological distribution	US scan findings ($n=30$)					CT scan findings ($n=19$)				
	None	Right	Left	Bilateral	Positive US scan	None	Right	Left	Bilateral	Positive CT scan
Hemiparesis										
right	1	–	4	–	4/5	3	–	1	1	2/5
Left	1	1	–	–	1/2	1	–	–	–	0/1
Diplegia	6	–	1	–	1/7	3	–	–	–	0/3
Triplegia										
right	2	–	–	2	2/4	2	–	–	1	1/3
left	1	–	–	1	1/2	2	–	–	–	0/2
Quadriplegia	1	–	–	–	0/1	2	–	–	–	0/2
Symptomatic total	12	1	5	3	9/21	13	0	1	2	3/16
Asymptomatic (normal) total	7	–	–	2	2/9	2	–	–	1	1/3
Combined total	19	1	5	5	11/30	15	0	1	3	4/19

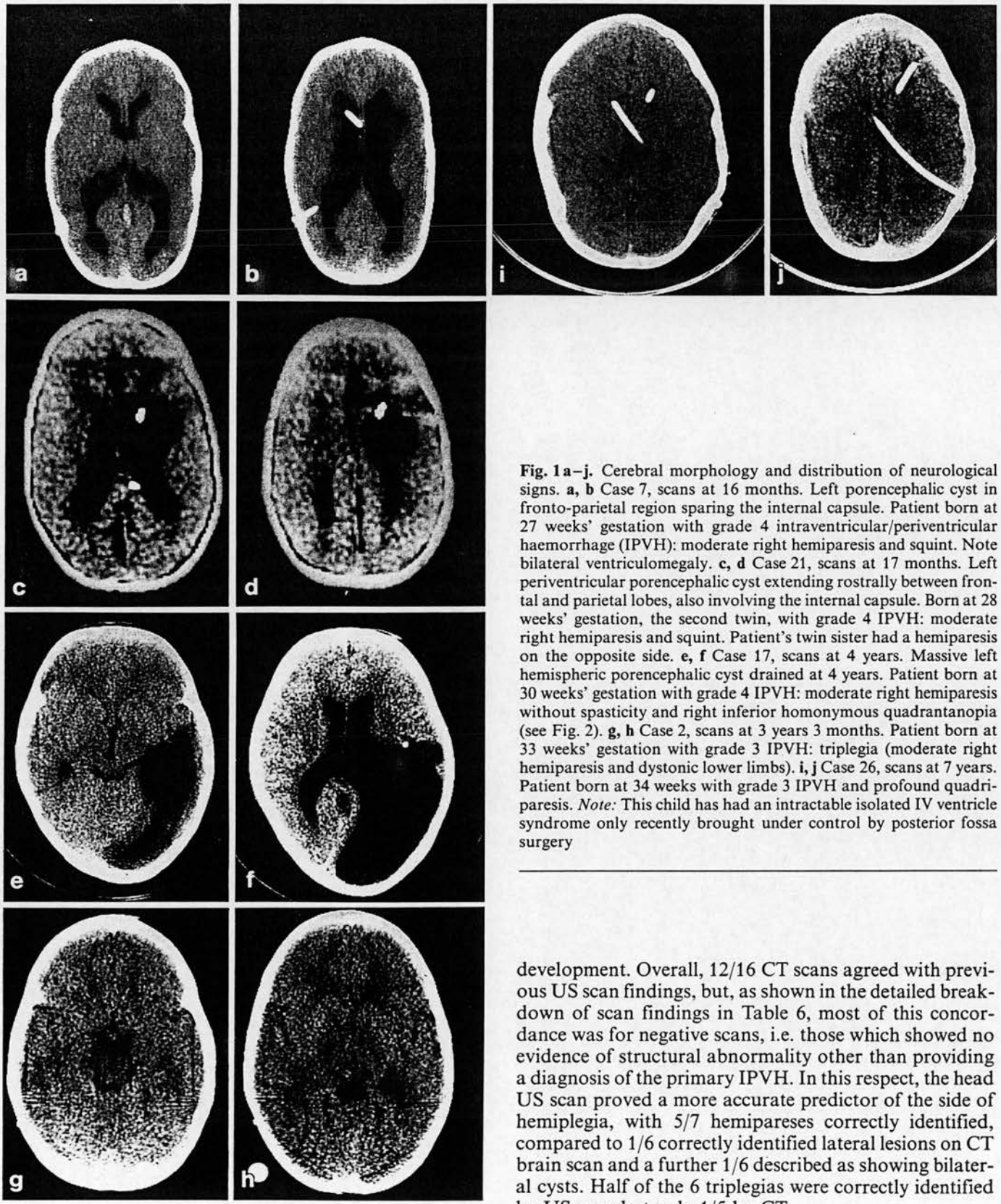

Fig. 1a–j. Cerebral morphology and distribution of neurological signs. **a, b** Case 7, scans at 16 months. Left porencephalic cyst in fronto-parietal region sparing the internal capsule. Patient born at 27 weeks' gestation with grade 4 intraventricular/periventricular haemorrhage (IPVH): moderate right hemiparesis and squint. Note bilateral ventriculomegaly. **c, d** Case 21, scans at 17 months. Left periventricular porencephalic cyst extending rostrally between frontal and parietal lobes, also involving the internal capsule. Born at 28 weeks' gestation, the second twin, with grade 4 IPVH: moderate right hemiparesis and squint. Patient's twin sister had a hemiparesis on the opposite side. **e, f** Case 17, scans at 4 years. Massive left hemispheric porencephalic cyst drained at 4 years. Patient born at 30 weeks' gestation with grade 4 IPVH: moderate right hemiparesis without spasticity and right inferior homonymous quadrantanopia (see Fig. 2). **g, h** Case 2, scans at 3 years 3 months. Patient born at 33 weeks' gestation with grade 3 IPVH: triplegia (moderate right hemiparesis and dystonic lower limbs). **i, j** Case 26, scans at 7 years. Patient born at 34 weeks with grade 3 IPVH and profound quadriparesis. *Note:* This child has had an intractable isolated IV ventricle syndrome only recently brought under control by posterior fossa surgery

development. Overall, 12/16 CT scans agreed with previous US scan findings, but, as shown in the detailed breakdown of scan findings in Table 6, most of this concordance was for negative scans, i.e. those which showed no evidence of structural abnormality other than providing a diagnosis of the primary IPVH. In this respect, the head US scan proved a more accurate predictor of the side of hemiplegia, with 5/7 hemipareses correctly identified, compared to 1/6 correctly identified lateral lesions on CT brain scan and a further 1/6 described as showing bilateral cysts. Half of the 6 triplegias were correctly identified by US scan but only 1/5 by CT.

The differences may be accounted for by differences in timing of the imaging, since in general US scans were performed earlier than the CT scans. Another important factor relates to the technology available for imaging before 1985.

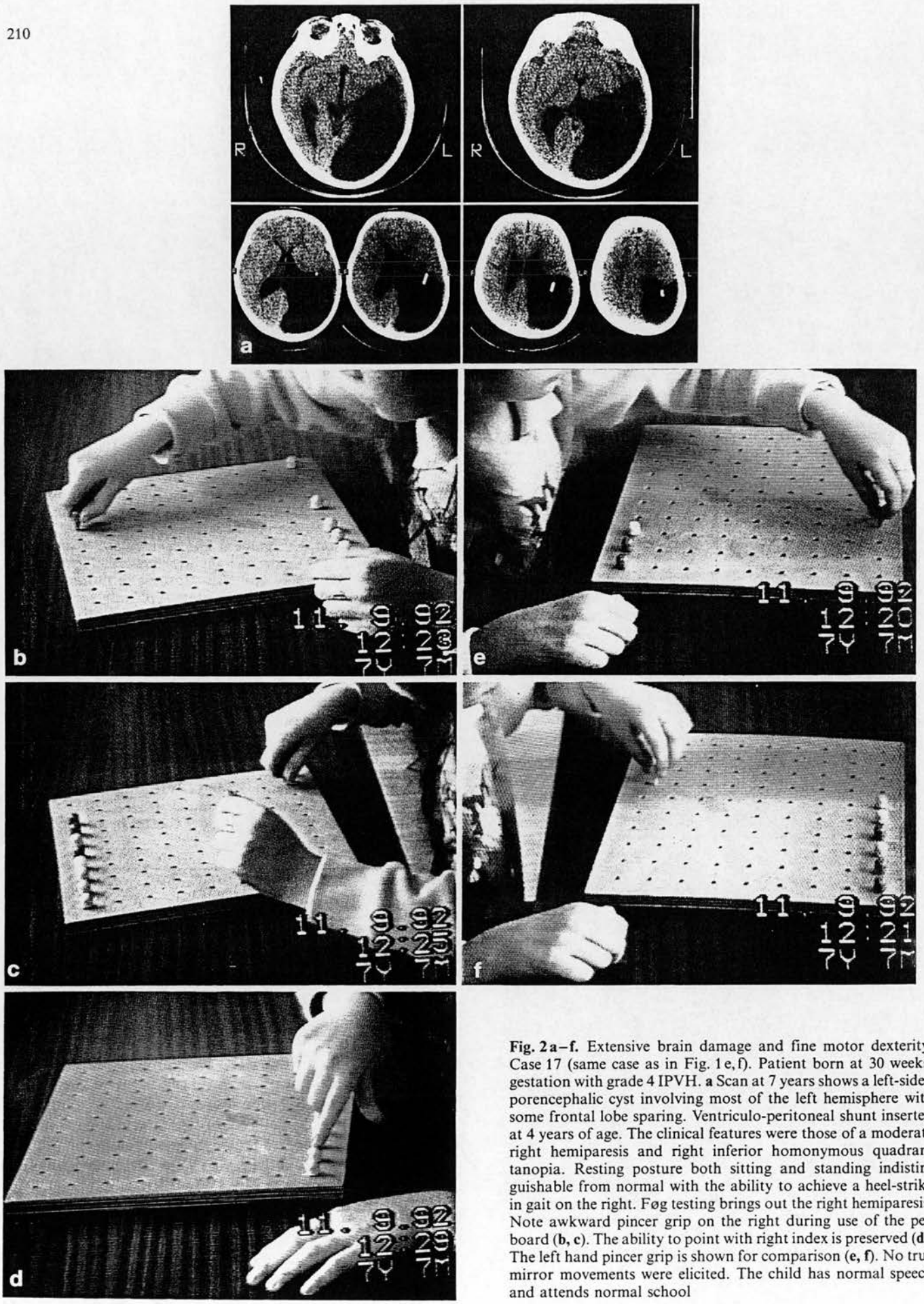

Fig. 2a–f. Extensive brain damage and fine motor dexterity. Case 17 (same case as in Fig. 1e, f). Patient born at 30 weeks' gestation with grade 4 IPVH. **a** Scan at 7 years shows a left-sided porencephalic cyst involving most of the left hemisphere with some frontal lobe sparing. Ventriculo-peritoneal shunt inserted at 4 years of age. The clinical features were those of a moderate right hemiparesis and right inferior homonymous quadrantanopia. Resting posture both sitting and standing indistinguishable from normal with the ability to achieve a heel-strike in gait on the right. Føg testing brings out the right hemiparesis. Note awkward pincer grip on the right during use of the peg board (**b, c**). The ability to point with right index is preserved (**d**). The left hand pincer grip is shown for comparison (**e, f**). No true mirror movements were elicited. The child has normal speech and attends normal school

Table 7. Accuracy of prediction of neurological impairment by US scan

Patient outcome	US scan		Total
	Negative	Positive	
Asymptomatic	7	2	9
Symptomatic	12	9	21
Total	19	11	30

Table 8. Accuracy of prediction of neurological impairment by CT scan

Patient outcome	CT scan		Total
	Negative	Positive	
Asymptomatic	2	1	3
Symptomatic	13	3	16
Total	15	4	19

Table 9. Concordance of CT scans with previous US scans[a]

Topographical neurological distribution	CT/US
Normal	2/2
Hemiparesis	
right	2/4
left	1/1
Diplegia	3/3
Triplegia	
right	2/3
left	1/2
Quadriplegia	1/1
Total concordance	12/16

[a] 4 CT scans negative when previous US scans had been positive. No cases of positive CT scan with negative US scan

Discussion

The clinical evaluation of children who have suffered a grade 3 or 4 IPVH reveals a wide heterogeneity of neuromotor syndromes, with great variations in tone abnormality and functional neuromotor impairment.

Comparison with previous reports

Past studies were limited to autopsy surveys of mainly neonatal deaths until the late 1970s, i.e. the babies investigated had not yet evolved any of the neurological syndromes which emerge with growth and maturation. Since Papille's original grading of the CT findings following IPVH in 1978 [12], there have been many live follow-up studies comprising data on all grades of IPVH. A number of different classifications of IPVH have been used, though the most commonly employed have all been variations on the Papille approach. Some studies have concentrated on producing a classification of impairments from normal through to multiple handicaps [13], or a quotient of impairment. [6]. Relatively few studies have looked at the pattern of neurological damage in relation to IPVH. One early study spanning all grades of CT-proven IPVH in 15 children showed at a 24-month follow-up that 6/15 were normal, 6/15 mildly and 3/15 moderately to severely impaired [8]. Impairments in this study referred to sensorineural deficits, speech immaturities as well as developmental motor delays and hard neurological signs. It is interesting that the 12/15 children with no or only mild impairments had suffered only grade 1 or 2 haemorrhages and had no motor neurological signs, whereas the 3/15 cases with moderate to severe impairment all had "spastic hemipareses" and had sustained grade 4 IPVH. This suggested an almost certain adverse outcome with grade 4 IPVH. A subsequent study, involving an 18-month corrected age screening of 228 mechanically ventilated low-birth-weight infants (<1750 g) with or without IPVH [9] demonstrated that the IPVH found in 51/228 screened neonates was associated with motor abnormalities regardless of anatomical location of the bleed, whether unilateral or bilateral germinal matrix or lateral ventricular haemorrhage: *none of the 5/51 cases with parenchymal haemorrhage developed signs of spastic CP*. The authors found that ventriculomegaly (including early ventriculomegaly in the first 7–13 days) in 15/51 IPVH cases carried a five-fold increased risk of spasticity and delayed walking together with a three-fold increased risk of hypertonus and hyper-reflexia, irrespective of gestational age. This association between ventriculomegaly and adverse outcome is supported by the earlier prospective cranial US findings of Stewart et al. [18]. The fact that only 3/15 of the ventriculomegaly cases required shunt operations casts doubt on whether medical or surgical intervention could have altered the course of such a progression towards an adverse outcome. Furthermore, the authors were unable to differentiate clearly between ventriculomegaly due to atrophy, gliosis or raised intracranial pressure, nor did they specifically look for periventricular echodensities or cysts.

We have previously described our functional outcome grades in terms of the raised intracranial pressure data prior to ventriculo-peritoneal (VP) shunting. Only in 7/33 cases were pressures not recorded, so that in the remaining 26/33 there were no cases of "normal pressure hydrocephalus" [11]. We were also able to demonstrate a worse outcome in children with more than 4 VP shunt revisions; however, the issue of what causes the damage – whether the primary IPVH, the ensuing ventriculomegaly, the shunt complications or indeed associcated factors such as coexisting ischaemic changes as a result of cerebral vasomotor instability in the perinatal period – remains unresolved, and we suspect that many factors contribute to the final picture. Cooke [5] reviewed 12 patients who underwent shunting for IPVH. Of 9/12 with grade 4 IPVH, 4/9 developed hemiparesis, 2/9 diplegia and 1/9 quadriplegia; spasticity was the commonest tone abnormality, affecting 3/9 cases. A porencepahlic cyst was as-

sociated with normal development in 1/7 cases, clumsiness in 1/7, hemiplegia in 3/7, diplegia in 1/7, and quadriplegia with superimposed hemiplegia in 1/7. This again demonstrates that conspicuous morphological changes on cranial imaging need not result in overt neurological signs, and in his study Cooke attributed the adverse outcome not to parenchymal changes but to fits in the 1st weeks of life, an association not borne out by our previous report. Another factor raised by Cooke was the better outcome from frequent ventricular taps prior to VP shunting, but this has been superseded by the recent European Multicentre Trial which failed to demonstrate a benefit from frequent ventricular tapping [19].

Catto-Smith and colleagues [4] reviewed 20 survivors with IPVH, of which 3/20 had an intracerebral bleed. Of these, 1/3 had no evidence of CP but was blind due to retrolental fibroplasia and severely handicapped, 1/3 was hemiparetic and 1/3 quadriplegic the latter two with mild and severe handicaps respectively. Of 8/20 cases with germinal layer bleeds (grade 1 IPVH), 2/8 were classified as being moderately handicapped because of a mental developmental quotient (MDQ) <80 and although one case also had epilepsy, neither had CP. Intraventricular haemorrhage (grades 2 and 3) occurred in the remaining 9/20 cases and resulted in hydrocephalus but no functional handicap in 1/9, mild-to-moderate handicap without CP in 2/9, and hemiplegia with mild handicap in 1/9 or quadriplegia with severe handicap in 2/9 cases: the remaining 3/9 cases of IVH were asymptomatic.

For these small numbers handicap may not relate to the presence of CP (non-progressive motor deficit), and CP may exist with only mild handicaps, yet Catto-Smith and colleagues concluded that "extreme pessimism is warranted" in the face of severe IPVH. The most recent 5-year follow-up of 25 cases of grade 3 and 4 IPVH [20] advocates a viewpoint of "guarded optimism" since only 6/25 had evidence of neurological impairment, though the exact nature of these impairments is not stated. The same study showed no influence of socioeconomic background on motor outcome, but showed that *all survivors of premature birth*, whether or not they had suffered from IPVH, had subtle neurodevelopmental changes on screening in the absence of frank neurological impairments. Although, of course, severe neurological disorders may result from the severest forms of IPVH, the studies quoted in detail show conclusively that there is a lack of an absolute correlation between grade of IPVH, volume and distribution of morphological changes on cranial imaging and the presence or absence of neurological signs. Furthermore, there have been no studies comparing the neurological features following IPVH and those of Little's diplegia of low birth weight or prematurity, so that it has been difficult on epidemiological grounds to determine which of these two separate conditions is contributing to the continuing prevalence of CP of 2–2.5/1000 live births.

Our study has looked at 33 cases of the severest grades of IPVH and analysed the neurological patterns in terms of topographical neurological distribution (TND) of signs, disordered tone and functional neuromotor grade (FNG) of disability.

Topographical neurological distribution

The overall picture is that the sequelae of the severest forms of IPVH do not simply resemble the "encephalopathy of prematurity" or Little's disease, since 10/33 patients were normal, leaving 23/33 with abnormal signs. Only 6/33 exhibited a diplegia, and in only 5/6 of these was there some resemblance to Little's diplegia of prematurity (though none were spastic), with delayed gross motor milestones and dystonic lower limbs; the remainder had hemiparesis (8/23), triplegia (6/23) and quadriplegia (3/23) respectively. We have found that (leaving out the normal and diplegic groups) the asymmetrical injury of grade 4 IPVH resulted in hemiplegia or triplegia, whereas grade 3 IPVH produced all 3 quadriplegic children. Since the severity of the initial haemorrhagic insult is judged to have been similar in all cases, some of these differences might be attributable to the varying circumstances of pregnancy, labour or neonatal course, but our previous findings were unable to confirm this [11]. Of the 8 children with hemiparesis, 2/8 were twin sisters born at 28 weeks' gestation with opposite hemisphere affected, producing a right and a left hemisyndrome respectively. This topographical neurological pattern for the twins must be related to some intrinsic common property of being twins such that the IPVH injury occurred in a mirror pattern. For the remaining children, the differences in neurological distribution can only be explained in terms of coexisting vasomotor instability causing ischaemia at the time of the injury or the complications of ventriculomegaly as outlined in our pathogenetic scheme for neurological damage [11]. Such subtle parenchymal changes would not necessarily show up on cranial US scan or CT imaging.

Topographical neurological distribution and functional outcome

As in other studies [7], the diplegic children did better on functional neuromotor assessments than purely hemiparetic children, who in turn did better than triplegic children. Quadriplegic children all had profound functional deficits.

Tone and outcome

The most common abnormality of tone was dystonia, which affected 11/23 cases with abnormal neurological signs (occurring alone in 8/23 or with hypotonia in 3/23 cases). Tone could be *normal* in the presence of moderate functional neuromotor impairment, as in the case of 7/8 of the hemiparetic children. From this small study of a specific group, the finding of ataxia/hypotonia tended to confer a slightly better functional outcome than a dystonic picture, which in turn matched a more favourable outcome than a mixed (hypotonic/dystonic) pattern of tone. Spasticity was rare. It is not possible to comment on or compare spasticity with function since spasticity was noted in only 1/33 cases. This single spastic case with a

quadriplegic distribution of neurological signs did have a profound functional impairment, whereas the quadriplegic child with dystonia has since developed sufficiently to push himself around in a wheelchair.

Imaging and prognosis

The US scan or CT brain scans proved poor predictors of neurological impairment except in the case of the hemipareses, for which the US scan but not the early CT scan proved accurate in 5/7 cases. Neither for the diplegias nor for the most severely affected children with quadriplegia did US or CT scanning provide any prognostic information. These findings contrast with those of Stewart et al. [18], who were able to place 342 at-risk neonates into a large group with a low risk and a small group with a high risk of neurodevelopmental abnormality, on the basis of the presence or absence on prospective US scanning of IPVH and parenchymal echodensities in the 1st week of life or subsequent ventricular dilatation or cerebral atrophy.

A recent study [10] which involved the follow-up of 155 survivors out of 200 very-low-birth-weight children showed that none of the 12/155 with CP had sustained a grade 4 IPVH. Of these 12/155 with CP, 3/12 had grade 3 IPVH and cystic periventricular leucomalacia (CPVL), which was associated with a hemiplegia in 2/3 and quadriplegia in 1/3 cases. By contrast, one child with a normal US scan had quadriparesis and another child with grade 1 IPVH had a diplegia. Grade 2 IPVH with CPVL was recorded in 5/12 of the CP group, which took the form of hemiplegia in 2/5, diplegia in 1/5 and quadriplegia and double hemiplegia in the remaining 2/5 grade 2 cases. A comparison of the test of motor impairment (TOMI) between these 155 survivors and 144 control children at the age of 5 years was unable to find differences between children in the overall score according to a classification of the US scan findings into normal, prolonged flare (PF), germinal matrix haemorrhage-intraventricular haemorrhage (GMH-IVH) without PF or GMH-IVH with PF. However, a weak statistically significant finding of poorer manual dexterity ($p<0.03$) was found in the 9/155 GMH-IVH+PF US scan group compared with the other US scan groups [10]. Although there was a weak association between the manual dexterity subscore of the TOMI and GMH-IVH with prolonged flare on US scan, this association was strongly influenced by birth weight, leaving the authors to speculate on the impact of nutrition and non-ischaemic damage from unrecorded episodes of hypoglycaemia on the scan-negative and scan-positive children. A similar association between poor neuromotor competence and low birth weight which was not attributable to overt disability has been found in the Scottish low birth weight study [15] of 636 children weighing less than 1750 g assessed at 4.5 years, but only for the subtasks of the TOMI related to bead threading, one leg balance and jumping over a cord.

The size and distribution of morphological changes on CT scanning does not necessarily equate with the severity of impairment as recently shown in 110 hemiparetic children [21]. These findings suggest that great caution must be exercised in taking decisions on the basis of brain imaging regarding ultimate prognosis or the predicted quality of life of an infant with severe IPVH. The corollary of being unduly optimistic because imaging has failed to indicate a conspicuous lesion should also be resisted. Our results show that ventriculomegaly in the first few weeks of life need not be associated with a poor prognosis in up to a third of all cases of severe of IPVH.

Conclusion

Grades 3 and 4 IPVH produced no neurological impairment in a third of our cases. Classical diplegia of prematurity represented only 6/23 of the cases with neurological signs and was equally distributed between the grade 3 and grade 4 IPVH groups. An asymmetrical injury (grade 4 IPVH) need not carry a worse prognosis than a symmetrical insult (grade 3 IPVH). The commonest impairment was hemiparesis in 8/23 children, and 7/8 of these had sustained a grade 4 IPVH, as had 5/6 of the triplegic cases, whereas all 3 quadriplegic cases belonged to the grade 3 IPVH group. The majority of hemiparetic children (7/8) had no tone abnormality, which is at variance with the notion that the majority of hemiparetic children are spastic. The children with diplegia were dystonic in 5/6 cases or had mixed dystonia/hypotonia in 1/6 cases. Of those with triplegia, 4/6 were dystonic, 1/6 ataxic/hypotonic and 1/6 dystonic/hypotonic. Only one child with quadriplegia had overt spasticity, the other 2/3 quadriplegic children having either dystonia or dystonia/hypotonia. This suggests that spasticity is not the commonest form of tone disturbance in this group of hemiplegic or diplegic children and should lead to a re-evaluation of the prevalence of "spasticity" among all cerebral palsied children. Cranial imaging restricted to US or CT scans is a relatively poor prognostic indicator of outcome when applied to the most severe cases of IPVH.

Most importantly, a clear distinction should be made between the features of the classical "encephalopathy of prematurity" known as Little's disease (which occurs in preterm infants without a conspicuous acute neurological illness in the neonatal period), which only produces a diplegia, and the neurological syndromes following IPVH, if trends on the prevalence of cerebral palsy are to be correctly interpreted and the success of neonatal care properly evaluated.

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PERIPHERAL AND CENTRAL MECHANISMS OF HINDFOOT EQUINUS IN CHILDHOOD HEMIPLEGIA

J-P. Lin
J. K. Brown

The childhood hemiplegias are now the most common of the cerebral palsy syndromes in children born at term, and the second most common among those born preterm (Hagberg *et al.* 1989). 90 per cent are congenital, the majority of which are judged to be of prenatal origin among term infants (Wiklund and Uvebrant 1991). Even allowing for the survival of larger numbers of preterm infants, which would alter the relative hemiplegia: diplegia ratio within the cerebral palsy population, or for changes in birthrate, the number of new hemiplegic children is unlikely to fall.

The clinical manifestations of hemiplegia are: loss of distal fine-motor dexterity and power, with or without limb atrophy; increased reflex excitability, including clonus, disturbances of gait, muscle tone and posture; trophic and vasomotor changes; and deformity of the upper and lower limbs. Children with hemiplegia usually walk unaided between 18 and 20 months (Crothers and Paine 1959, Bleck 1975) and, if seizure-free, most are able to attend normal schools. Many are athletic, taking part in sports, swimming and cycling, but most will never develop the heel-strike (Fig. 1) which is the hallmark of the mature pattern of walking, normally acquired between the second and fourth years of life (Sutherland *et al.* 1980, Leonard *et al.* 1991). This deviant pattern of walking appears to be related to the development of pes equinus. The equinus foot may variably affect walking and running, and may require regular physiotherapy, passive splinting, expensive footwear or surgery; yet a detailed understanding of the mechanisms of equinus is lacking (Table I).

The clinical impression of a lack of correlation between the distribution and size of the brain lesion and the abnormalities of gait and posture has recently been confirmed by the study of 111 CT scans from hemiplegic children (Wiklund and Uvebrant 1991), which showed no association between specific morphological patterns or the extent of brain damage and gait disturbance. Whereas *arm*-dominant hemiplegia appeared to be more often associated with what the authors termed 'maldevelopment' and 'cortical/subcortical atrophy', *leg*-dominant hemiplegia was more likely to be associated with a normal scan. Equinus associated with hemiplegia is unpredictable, its mechanisms are obscure and its treatment insecure.

Neurophysiological methods of examining and assessing the lower limb in hemiplegia by comparing the affected with the 'normal' side have been described (Brown *et al.* 1991). The present study was undertaken to evaluate the possible contribution to hind-foot equinus

TABLE I
Possible mechanisms of hindfoot equinus

Short leg
Short tendon
Short muscle
Foot-drop
Stiff muscle
Tonic spasticity
Dystonia
Rigidity
Phasic toe-strike
Hemiplegic posture
Abnormal walking engram

TABLE II
Timing and aetiology of hemiplegia

Congenital/perinatal	
Silent infarcts	10
Asphyxia	4
Intra-uterine growth retardation	3
Preterm birth	3
Acquired	
Trauma	1
After by-pass for tetralogy of Fallot	1
After measles	1
Rasmussen syndrome	1
	24

of a number of readily measurable physical variables, using simple bedside techniques.

Patients

Twenty-four hemiplegic children attending the Cerebral Palsy Service at the Royal Hospital for Sick Children in Edinburgh were consecutively selected to take part in the study between January and June 1991. Only children aged five of over were included, so that they would be able to co-operate in the various examinations. All examinations took place in the presence of and with the consent of the parents. The population characteristics are detailed in Table II and in Appendix Table A. The balance of 20 children with congenital and four with acquired hemiplegia broadly reflects the composition of our clinic population. The ages spanned 6·2 to 17·7 (mean 9·9) years. Half of the 20 children in the congenital group were presumed to have silent infarcts; the remainder had evidence of perinatal asphyxia, intra-uterine growth retardation and preterm birth. Traumatic, vascular and infectious aetiologies for onset of hemiplegia, which account for 90 per cent of all acquired hemiplegias, are all represented in our four cases with acquired hemiplegia. There were 14 cases of right and 10 of left hemiplegia.

Method

Some of the possible factors contributing to hindfoot equinus are listed in Table I. The clinical variables assessed are listed in Table III.

Definition and measurement of equinus
Hindfoot equinus was defined in two ways: (1) gait equinus—*an obligate toe-strike maintained throughout stance in the gait cycle;* and (2) passive equinus—*a compliance difference of >10°/unit torque in passive dorsiflexion.* Compliance difference was measured with the child supine. Equal pressure was applied simultaneously to the soles of the feet: the difference in angular displacement at the ankle joint represented the compliance difference in degrees/unit torque (Fig. 1).

Anthropometry

LEG LENGTH

The leg-length difference between the normal and affected limb was recorded in centimetres from the anterior superior iliac spine to the medial malleolus, with the pelvis straight.

ANKLE-JOINT RANGE

Ankle-joint ranges (in degrees) were obtained for normal and affected limbs, and the difference was recorded. Joint angles were assessed by measuring the angle of the sole of the foot in relation to the shaft of the tibia: neutral was defined as the position when the sole is perpendicular to the tibia, and the angles of dorsiflexion and plantarflexion were measured relative to this. The position of resting equinus was measured for normal and affected sides in all cases with the child supine.

MUSCLE EXTENSIBILITÉ

Extensibilité difference was measured in degrees. The term 'extensibilité' was coined by André-Thomas and de Ajuriaguerra (1949) to define that portion of the total joint-range corresponding to

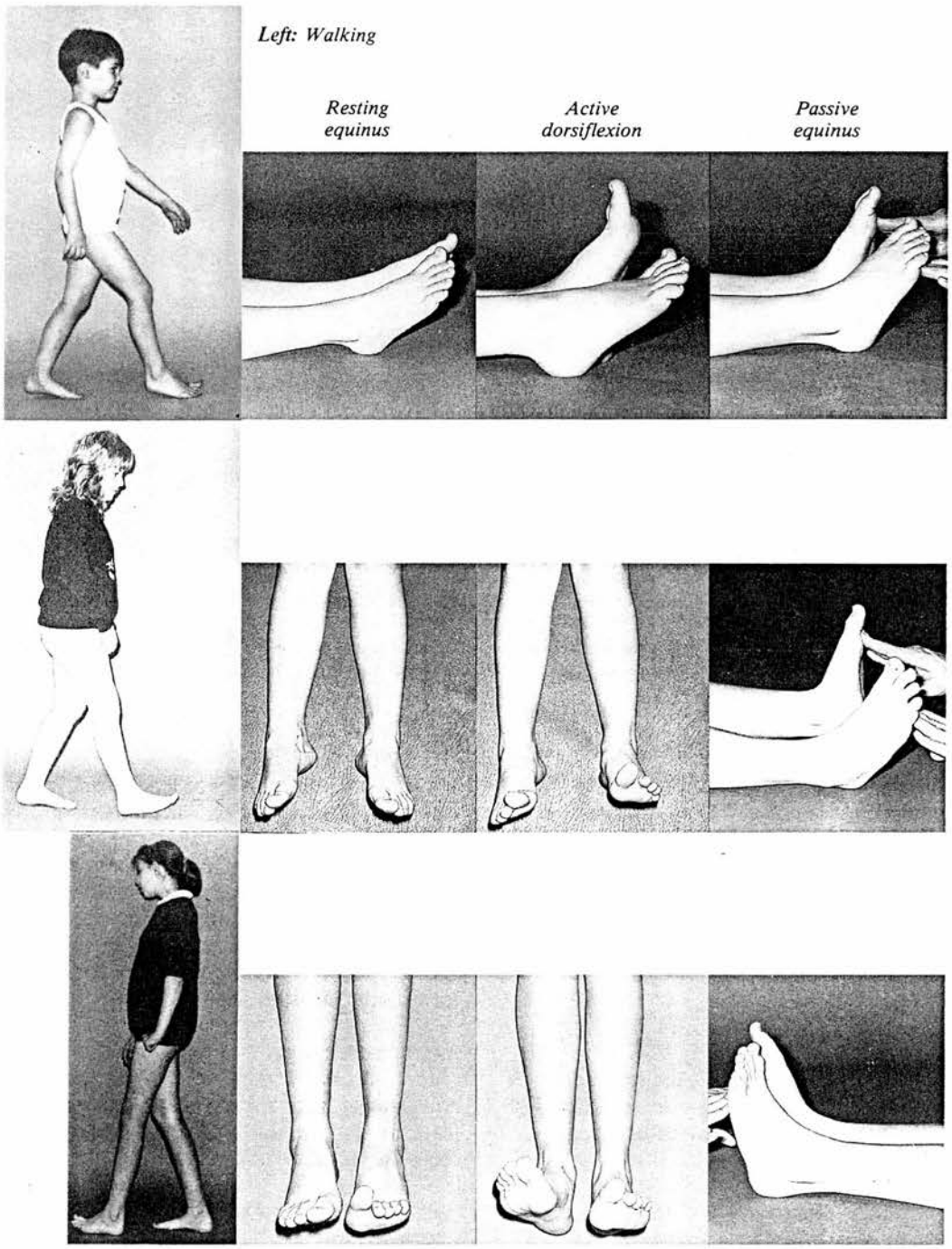

Fig. 1. Gait equinus, resting equinus and passive equinus in childhood hemiplegia. (Top) *Case 17: six-year-old boy with congenital right hemiplegia and gait equinus (toe-strike).* (Centre) *Case 6: nine-year-old girl with congenital right hemiplegia and heel-strike.* (Bottom) *Case 23: 12-year-old girl with congenital left hemidystonia due to discrete right pallidal infarct, showing normal heel-strike. Resting equinus for each case with patient supine is shown to be similar for normal and affected side, irrespective of power ranges or muscle tone. Active dorsiflexion brings out weakness of affected limb for case 17. All three cases had passive equinus, a compliance difference of >10°/unit torque: equal pressure is applied to soles of feet and difference in angular displacement is compliance difference in °/unit torque. For whole group, gait equinus could occur in absence of weakness and heel-strike with short, clinically spastic legs.*

TABLE III
Clinically assessed variables

Variable	*Units*
Leg-length difference	Centimetres
Ankle-joint range	Degrees
Extensibilité*	Degrees
Compliance difference	Degrees/unit torque
Gait score	Grade 1 = heel-strike Grade 2 = plantar-strike Grade 3 = toe-heel strike and back-knee Grade 4 = obligate toe-strike
MRC power grade†	Grade 0 = no contraction Grade 1 = flicker of contraction Grade 2 = movement, gravity eliminated Grade 3 = movement against gravity Grade 4 = movement against resistance Grade 5 = normal power
Distal dexterity of toes	Frequency of alternating flexion/extension in Hz
Clinical muscle tone	Phasic spasticity = resistance to rapid stretch Tonic Spasticity = resistance to slow stretch Dystonia = widely fluctuating co-contraction Rigidity = uniform, unvarying co-contraction

*That portion of total joint-range attributable to muscle lengthening.
†Medical Research Council (1976).

muscle lengthening. Joint angles were assessed visually. The tension corresponding to muscle lengthening was gauged by the examiner. The angle from the first onset of tension in the tendon until the onset of a further steep rise in tension was measured, and this represented the extensiblité of the muscle in degrees.

SIDE OF HEMIPLEGIA

The side of the hemiplegia was noted in each case to determine whether laterality influences the development of equinus.

GAIT SCORES

Gait was scored on a four-point scale, which is detailed in Table III.

POWER GRADES

The Medical Research Council muscle-power grades were used (MRC 1976). Muscles were tested isometrically. A muscle was said to have normal power if no difference could be found between affected and unaffected limbs. Power was tested with the child supine and, in the case of the plantarflexor and dorsiflexor muscle groups, with the knee fully extended. In each case the optimal maximum voluntary contraction sensed by the examiner was used as the final score, with no attempt at standardising the ankle-joint position, since the children positioned their ankle joints at the angle most comfortable for them.

MOTOR DEXTERITY

Fine-motor dexterity was determined by measuring the maximum frequency of alternating flexion and extension of the toes in Hertz.

DEFINITION AND ASSESSMENT OF MUSCLE TONE

Muscle tone was defined as the resistance felt as a limb is passively rotated about a joint at rest.

Phasic spasticity was used to describe the resistance to rapid stretch, and tonic spasticity to describe the resistance to slow stretch. Dystonia was used to describe wide fluctuations in tone due to co-contraction of muscles with changes of

TABLE IV
Differences between equinus and non-equinus groups as defined by gait and compliance difference (N = 24)

Variable	*Gait equinus (N = 9) Mean (SD)*	*No gait equinus (N = 15) Mean (SD)*	*Significance (test)*[1]	*Passive equinus (N = 16) Mean (SD)*	*No pass.equinus (N = 8) Mean (SD)*	*Significance (test)*[1]
Leg-length difference (cm)	2·0 (1·1)	1·8 (1·4)	NS *(t)*	2·2 (1·2)	1·1 (1·0)	$p<0·03$ *(t)*
Ankle-joint range difference (°)	12·5 (8·0)	8·0 (7·7)	NS *(t)*	12·5 (7·3)	2·9 (4·9)	$p<0·006$ *(t)*
Extensibilité difference (°)	10·0 (6·1)	8·7 (9·7)	NS *(t)*	12·2 (8·2)	3·1 (5·3)	$p<0·02$ *(t)*
Compliance difference (°/unit torque)[2]	23·9 (9·6)	14·5 (13·1)	NS *(t)*	—	—	—
Toe dexterity flexion/ extension (Hz)	2·1 (0·5)	2·4 (0·8)	NS *(t)*	2·3 (0·8)	2·2 (0·5)	NS *(t)*
Hemiside						
Right	6	8	NS (χ^2)	8	6	NS (χ^2)
Left	5	5		8	2	
MRC power scale[3]						
Plantarflexion (PF)	—	—	NS (W)	—	—	NS (W)
Dorsiflexion (DF)	—	—	NS (W)	—	—	NS (W)
Imbalance (PF-DF)	—	—	NS (W)	—	—	NS (W)
Clinical tone						
Phasic spasticity						
No	8	6	NS (χ^2)	8	6	NS (χ^2)
Yes	3	7		8	2	
Tonic spasticity						
No	6	4	NS (χ^2)	4	6	$p<0·02$ (χ^2)
Yes	5	9		12	2	
Hemidystonia						
No	8	14	NS (χ^2)	14	8	NS (χ^2)
Yes	1	1		2	0	

All differences calculated between normal and affected limb.
[1]NS = not significant; *t* = unpaired student *t* test; W = unpaired Wilcoxon rank sum test.
[2]No compliance differences given for passive equinus since it defines passive equinus.
[3]Medical Research Council (1976). No entries for power grades since these are ordinal numbers (see Fig. 2).

body position, and in response to non-specific afferent stimuli—such as conversation, noise and mental arithmetic—together with bizarre, sustained postures. Muscle tone was assessed clinically and graded according to whether it was normal or showed evidence of phasic, tonic or mixed phasic/tonic spasticity or hemidystonia.

Statistics

Differences between means were assessed using the unpaired Student *t* test. The χ^2 test was used to determine differences between groups, and the unpaired Wilcoxon rank sum test was used to analyse differences in ordinal grades of power between groups.

Results

Table IV summarises the main findings for all 24 cases of hemiplegia according to equinus and non-equinus categories. Figure 1 illustrates clinical differences between three hemiplegic children (cases 6, 17, 23) from the study group when walking at a self-selected speed, supine resting equinus of the normal and affected limbs, and active bilateral dorsiflexion and passive dorsiflexion by the examiner to elicit the compliance difference.

Goniometry

There were no cases of fixed equinus among the 24 children: all affected ankle-joints could be passively dorsiflexed beyond neutral. Over-all, there was a weak statistically significant reduction in ankle-joint range on the hemiplegic side compared with the normal limb ($p<0·03$), with a margin of error of $\pm 5°$. The children with right hemiplegia had significant differences in ankle-joint range ($p<0·006$) (Appendix Table B). Normal and affected limbs had similar angles of resting equinus with the child supine. The resting angle of the affected limb was not

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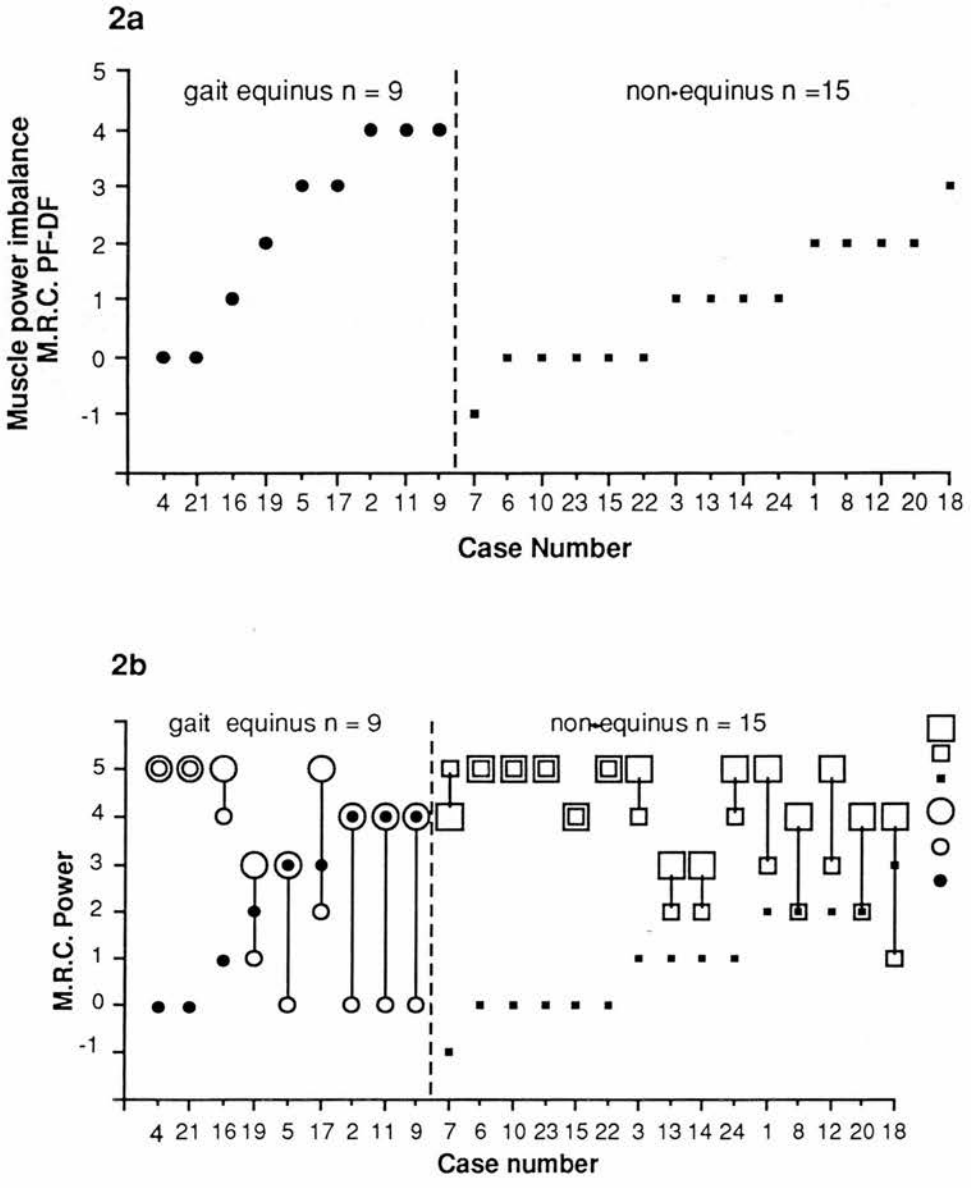


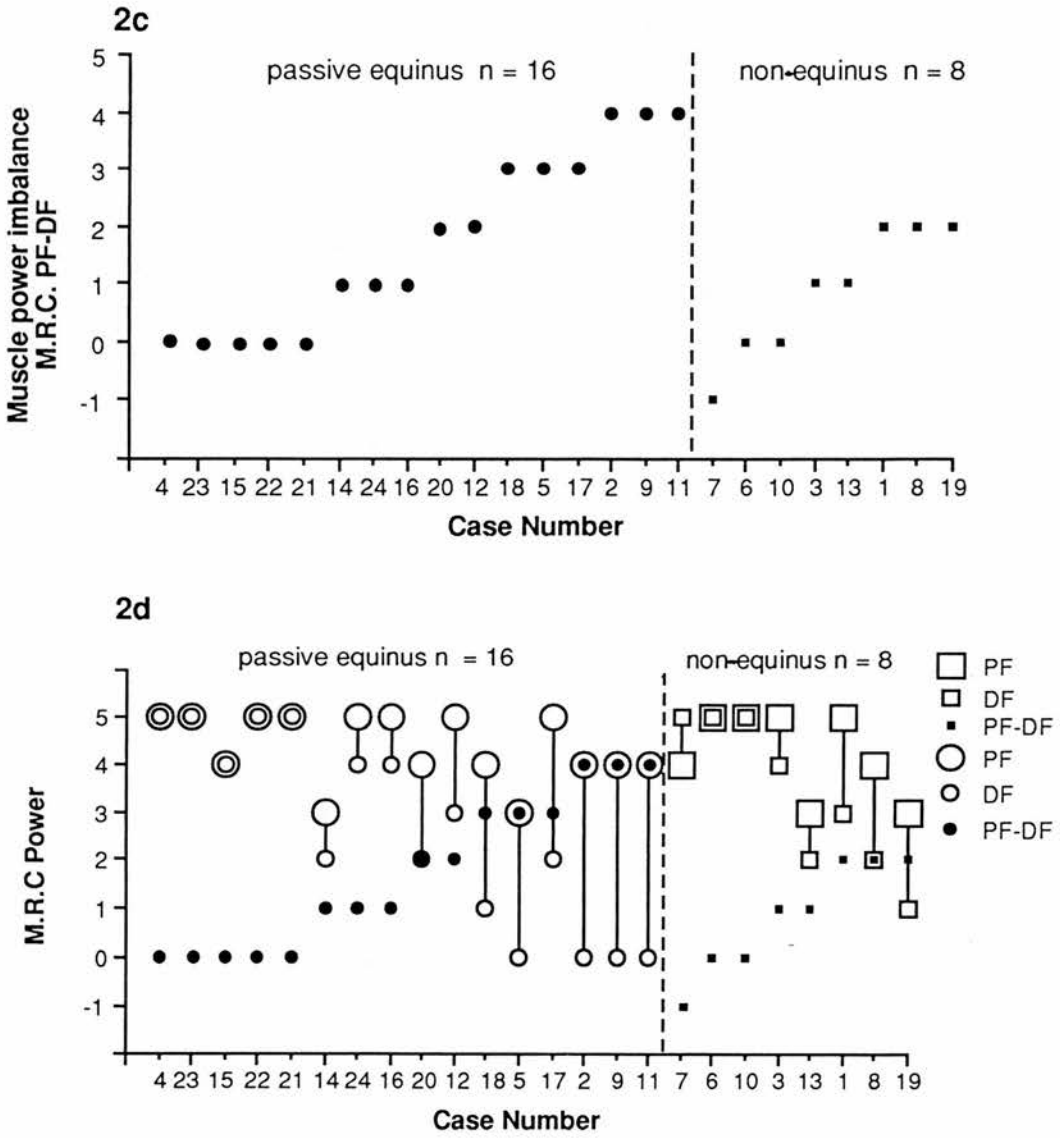
Fig. 2a-d. *Isometric muscle power and muscle imbalance at ankle. Case ranking according to increasing MRC muscle imbalance for gait equinus* (a) *and passive equinus* (c), *and their respective non-equinus groups. Actual MRC power grades are superimposed on same imbalance rankings for each case of gait equinus* (b) *and passive equinus* (d). *Note similarities in muscle imbalance and actual power values for all groups, irrespective of equinus status. Cases 4, 6, 10, 21, 22 and 23 have normal power. Case 7 has stronger dorsiflexors than plantarflexors, resulting in a 'negative' imbalance score.* Solid symbols = *imbalance scores;* large open symbols = *plantarflexor power;* small open symbols = *dorsiflexor power.*

helpful in predicting which hemiplegic limb would have an equinus gait.

The resting equinus in normal and affected limbs of those with left hemiplegia was significantly less than that of those with right hemiplegia, a finding for which there is no explanation. Details of goniometry findings are given in Appendix Table B.

Gait equinus

There were nine cases of gait equinus of all ages. Of the remaining 15 children, three had heel-strike, seven had plantar-



strike and five had toe-heel gait (see Fig. 1). A comparison of the clinically assessed variables between the groups with and without gait equinus showed no statistical differences in leg length, ankle-joint range, extensibilité, fine-motor dexterity, the side of hemiplegia, MRC power grading of plantarflexors and dorsiflexors or of muscle imbalance scores across the ankle joint, nor any difference in muscle tone at rest in the affected limbs for either group (Table IV). Although eight cases of gait equinus had compliance differences which satisfied the criteria for passive equinus, there was no over-all variation in compliance difference between the group with gait equinus and those with other gaits.

Passive equinus

In contrast, the 16 cases with passive equinus (a compliance difference of $>10°$/unit torque) had statistically shorter legs ($p<0{\cdot}03$), reduced ankle-joint ranges ($p<0{\cdot}006$) and reduced extensibilité ($p<0{\cdot}02$), together with a clinical impression of tonic spasticity ($p<0{\cdot}02$) of the affected limbs compared with the affected limbs of those with normal compliance. 12 of the 14 with

clinical evidence of tonic spasticity had criteria for passive equinus. There were no differences in fine-motor dexterity, the side of hemiplegia, or in the MRC power grading of the affected limbs between groups (Table IV).

MRC power grades

Figures 2a and 2c show the isometric power at the ankle in order of increasing imbalance for those with gait equinus, passive equinus and their respective non-equinus groups. Similar trends of muscle imbalance are apparent, irrespective of group category. The actual plantarflexor and dorsiflexor power values have been superimposed on the imbalance rankings in Figures 2b for gait equinus and 2d for passive equinus. There was an unexpected finding of apparently normal power in six cases, of whom cases 4 and 21 had both gait and passive equinus, cases 22 and 23 had passive equinus alone, and cases 6 and 10 had no equinus by either definition. The lack of weakness in cases 21 and 23 can be explained by both having a hemidystonic syndrome in which there was no muscle wasting; power of the anterior and posterior calf compartments was preserved, but functional impairment resulted from strong, involuntary co-contractions (subtraction weakness). Another singular finding was that of stronger power in the anterior rather than posterior compartment in case 7, which accounts for an imbalance score of −1 in Figure 2.

Discussion

With clinical experience, subtle differences in the clinical expression of brain damage can be gauged by structured observation of the hemiplegic child. Instrumentation is not required, and in fact may obscure important pattern differences. However, once these clinical differences have been elicited, more detailed study is necessary to understand their possible mechanisms.

In previous studies, hemiplegic children have been considered as a homogeneous group (Brown *et al.* 1987, 1991), the normal and affected limbs being compared using quantitative measurements of limb function. In contrast, the present study recognised the hemiplegias as being heterogeneous and attempted to identify factors that result in different signs and motor developments of the affected side by distinguishing children with and without a functional impairment, *i.e.* gait equinus. It also sought to determine whether an operational definition of hind-foot equinus, such as passive equinus, elicited during the clinical examination, correlates with functional impairment and other peripheral variables.

Gait equinus, leg length, ankle-joint range and extensibilité

Nine of the 24 children had obligate toe-strike and toe-stance. This contrasts with a study using foot-switch contact patterns, in which no equinus gait was found in the hemiplegic group (Csongradi *et al.* 1979), though only two hemiplegic limbs were observed. Heel-strike occurred in only three of our 24 cases, testifying to the over-all abnormality of gait on the affected side in hemiplegia.

It had been anticipated that gait equinus would be associated with limb atrophy on the basis of a compensatory equinus, but we could not confirm such a mechanism, despite a mean leg-length difference between normal and affected limbs of 1·9cm (SD 1·3cm, range 0 to 4cm). Children with one short leg but no neurological signs walked with a Trendelenburg gait, not compensatory hindfoot equinus; this is in accord with our findings in hemiplegia, although forefoot equinus may occur occasionally.

We could identify no association with other peripheral factors, such as reduced ankle-joint ranges or limited muscle extensibilité, which supports the theory that these peripheral changes do not cause gait equinus, but are the consequences of a common lesion.

Gait equinus, muscle power and the 'classical' power model of equinus

The lack of a clear correlation with dorisiflexor weakness, equivalent to central foot-drop, or muscle imbalance due to reciprocal inhibition at the ankle joint, was the most surprising result for the gait equinus group. Gait equinus is more complex than a simple, central, paralytic foot-drop (see Table IV, Figs. 2a, 2b).

Classical teaching on the mechanisms of deformity embraced four basic

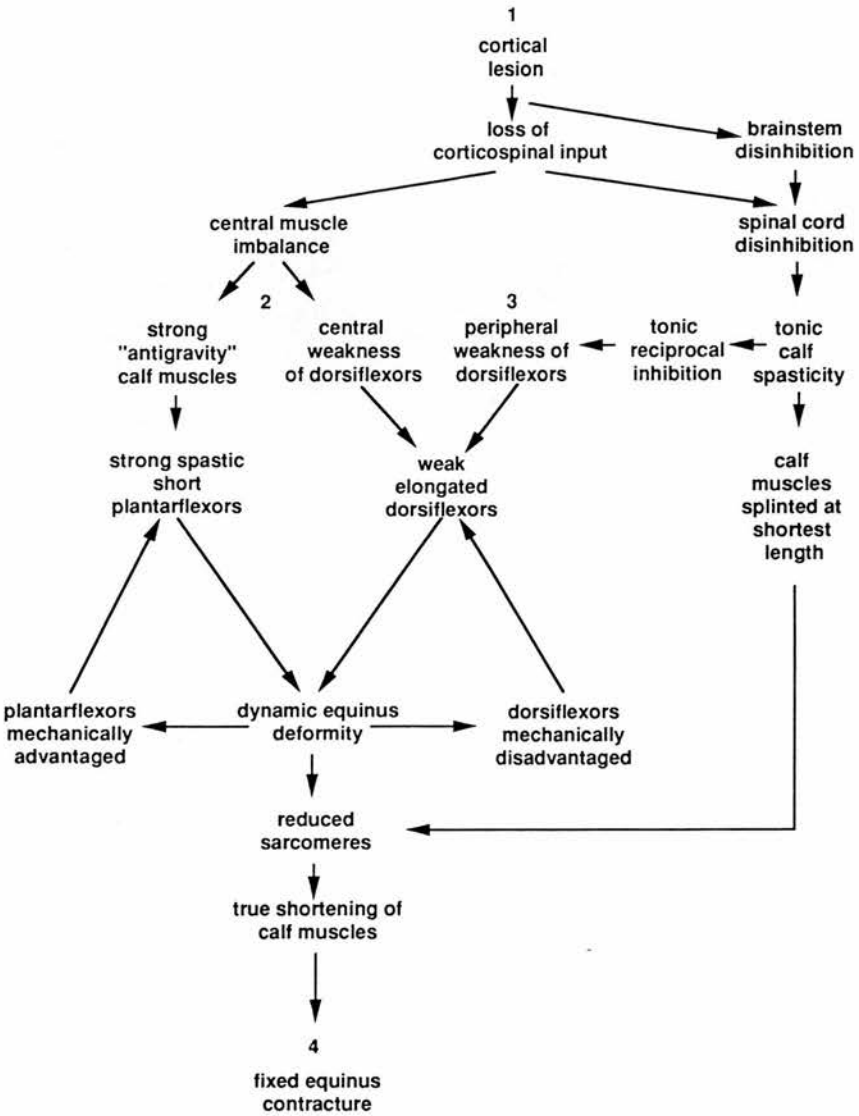


Fig. 3. Classical power model of equinus deformity. 1. *A cortical lesion results in central weakness and spasticity.* 2. *A central imbalance at ankle produces strong, 'antigravity' calf muscles and weak dorsiflexors.* 3. *Calf spasticity causes tonic reciprocal inhibition of dorsiflexors and peripheral weakness.* 1, 2 *and* 3 *combine to produce strong, spastic, short plantarflexors and weak elongated dorsiflexors, resulting in equinus posture. Equinus posture was thought to place calf muscles at mechanical advantage, based on belief that optimum power occurs at resting length of muscle. In equinus, dorsiflexors were thought to be at gross mechanical disadvantage. A vicious circle sustains equinus posture, resulting in eventual true shortening of calf muscles and fixed equinus contracture.* 4. *This scheme rests heavily on notion of weak dorsiflexors and 'strong' plantarflexors.*

premises to explain evolving muscle imbalance at the ankle joint (Fig. 3); these represented a synthesis of the known mechanisms of deformity following lower motor neuron lesions, *i.e.* polio and spina bifida, and the inferred pathophysiology of spastic states. This view of equinus is dominated by the concepts of muscle weakness and imbalance (Fig. 3), as opposed to abnormal muscle activation or temporal sequencing patterns (Fig. 4).

The first premise is that of central weakness, arising from reduced corticospinal input, with relative imbalance of

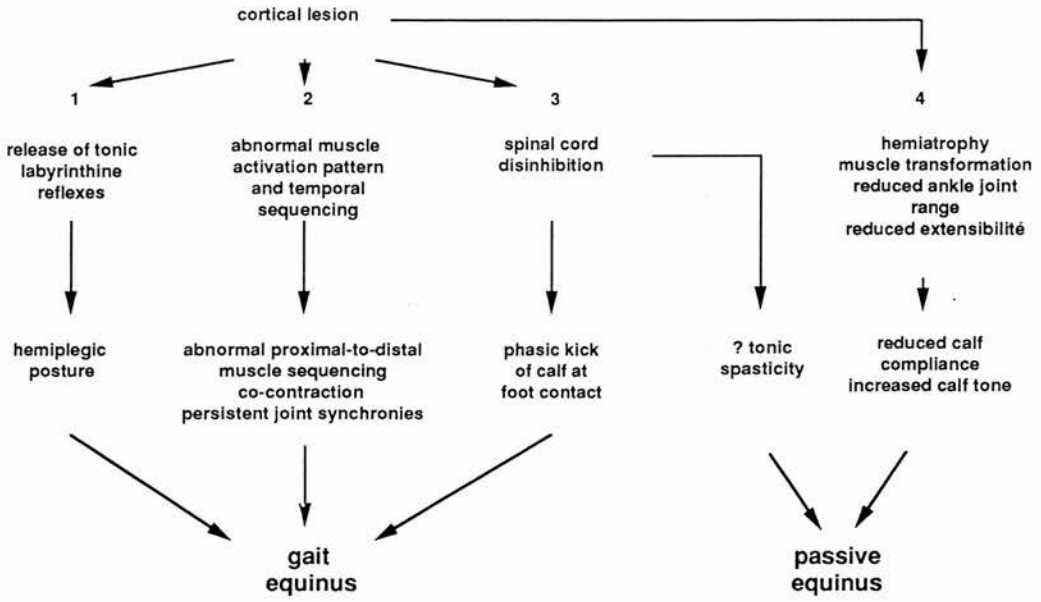


Fig. 4. Developmental model of equinus in hemiplegia.

the dorsiflexor to calf muscles. It should be remembered that normally there is a very great difference in torque between the powerful gastrocnemius-soleus complex and the anterior compartment muscles (Bleck 1987), leading many to wonder why normal individuals do not develop equinus on the basis of a normal physiological muscle imbalance. All our children had similar degrees of resting equinus on the normal and hemiplegic sides.

The second premise relates to spasticity of the calf, which was considered to cause tonic reciprocal inhibition of dorsiflexor agonist contraction, exacerbating the central dorsiflexor weakness by a peripheral spinal mechanism, leading to further muscle imbalance across the ankle joint. This peripheral dorsiflexor weakness is proportional to the severity of the spasticity. Sharrard (1974) taught that gait improved if the muscle imbalance was restored, and this could be achieved by weaknening the calf and removing the spasticity by unloading the stretch on the calf.

The third premise, an extension of the second, related to the observation that power in the dorsiflexors varies according to the position of the knee joint, the angle of which determines the tension, and hence spasticity, of the gastrocnemius muscle. The power of the dorsiflexors can thus be revealed if the knee joint is flexed, the calf muscles unloaded and the spasticity relieved (Silfverskiöld 1923). This led to differences in opinion among orthopaedic surgeons about whether power could be tested accurately in upper motor neuron syndromes in the presence of spasticity (Fulford 1984; personal communication).

The fourth premise was failure of muscle growth. Deformities arise much more commonly during the growing period. Normally the muscles on each side of a joint grow to accommodate bone growth. A muscle will grow longer and add sarcomeres if stretched, or shorten and remove sarcomeres if no opposition or forced lengthening occurs (Tardieu *et al.* 1988). Since the dorsiflexors are weak, there is no stretch on the calf muscles, which results in failure of normal growth, a reduction in the number of sarcomeres and a true shortening of the muscle, causing a fixed muscular contracture.

Central paresis with muscle imbalance, spasticity and reciprocal inhibition seemed logical and interdependent mechanisms for the production of a short muscle and a fixed contracture (see Fig. 3). This classical scheme can be

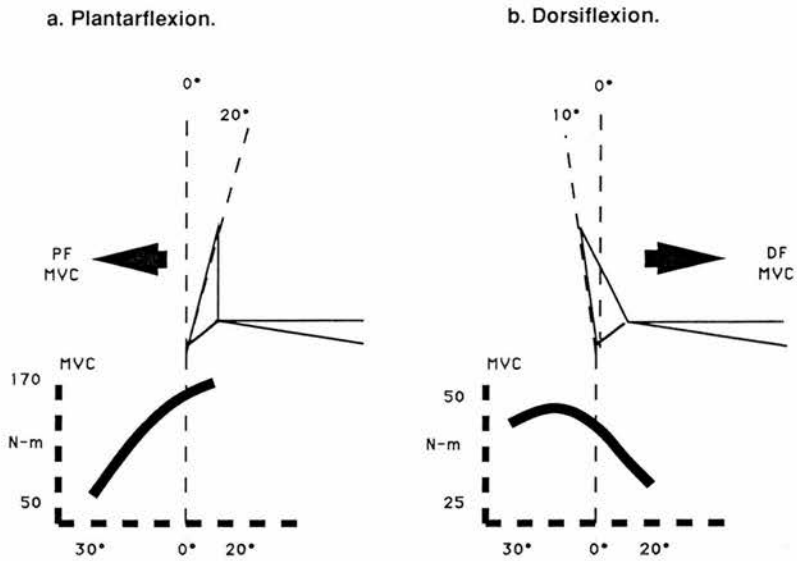


Fig. 5. Mechanics at ankle. Optimal maximum voluntary contraction (MVC) at ankle-joint (above) *and corresponding length-tension curves* (below): (a) *plantarflexor power (PF) is maximal at 20° dorsiflexion and weakest at 30° plantarflexion;* (b) *dorsiflexor power (DF) is maximal at 10° plantarflexion, rapidly decreasing beyond 5° dorsiflexion. Note different torque scales for MVC in* a *and* b. *This shows that plantarflexors are weakest in equinus and that equinus favours dorsiflexion (after Marsh* et al. *1981, Sale* et al. *1982 in adults).*

modified to take into account the non-neurogenic phenomenon of increased peripheral muscular stiffness, demonstrated by Foley (1961) to be independent of reflex activity.

In practice, the clinical assessment of power is confounded by the fact that there are optimal ankle-joint positions for dorsiflexor and plantarflexor maximum voluntary contractions (MVC) (Fig. 5), which do not correspond to the resting lengths of the dorsiflexors or plantarflexors (Marsh *et al.* 1981, Sale *et al.* 1982). The tibialis anterior is stronger at 30° of plantarflexion than in the neutral position, and plantarflexors are weaker with increasing plantarflexion. This finding suggests that the equinus posture favours the dorsiflexors to the detriment of plantarflexion, and that plantarflexors are weakest in equinus: a complete refutation of classical teaching.

These length-tension characteristics have implications for the assessment of muscle power of normal and cerebral-palsied children, as well as therapeutic implications. The MRC memorandum No. 45, 'Aids to the examination of the peripheral nervous system', shows the gastrocnemius being tested in a position of virtual maximum plantarflexion, *i.e.* a position of maximum weakness, while the tibialis anterior is assessed at the neutral position—positions now known to be sub-optimal for MVC. For most of the children in our study, the ankle joint was allowed to adjust to the position of optimum isometric MVC for that child when testing both plantar- and dorsiflexor power.

Since walking does not involve isometric contraction, extrapolation from static length-tension relations must be treated with caution. Nevertheless, newer evidence on ankle mechanics upsets older concepts of the pathophysiology of equinus, but accommodates the notion of abnormal central motor activation patterns.

Gait equinus and muscle tone

Gait equinus could not be explained in terms of increased muscle tone in the form of tonic spasticity as conventionally

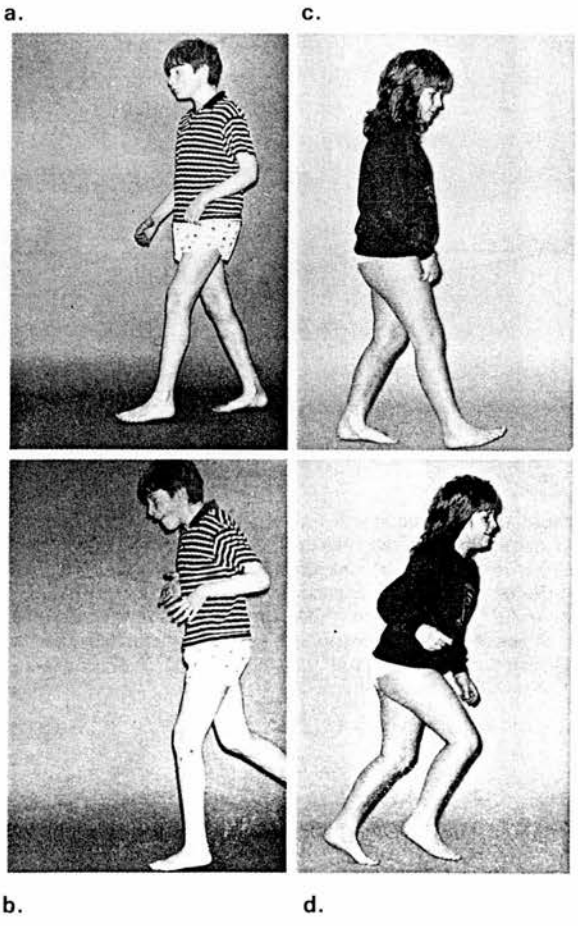


Fig. 6. Running and 'physiological' hemiplegic posture. Hemiplegic posture varies with activity on normal and affected sides. (a) *12-year-old boy with left hemiplegia and toe-heel gait (case 18):* (b) *note that running accentuates left hemiplegia posture and brings out right 'physiological' hemiplegic posture.* (c) *nine-year-old girl with right hemiplegia and heel-strike (case 6):* (d) *mature pattern of running, showing bilateral toe-strike with hemiplegic posturing of both upper limbs, accentuated on right.*

defined when clinically assessed at rest, or by inference from reduced compliance as defined above. Nor were we able to link the presence of phasic spasticity to gait equinus. All but one of the children with gait equinus had passive equinus, but only half of those with passive equinus had an equinus gait. This suggests that gait equinus is independent of reduced compliance.

Gait equinus and the hemiplegic posture

An alternative hypothesis to explain the development of gait equinus in accord with these findings is that gait equinus results from a superimposed hemiplegic posture. This posture is thought to emerge as a result of release from frontal-lobe cortical inhibition of tonic labyrinthine input to the spinal cord (Walshe 1923). It is reversed by holding the child upside down, and is not simply due to 'antigravity hypertonus', muscle imbalance or spasticity, since the hemiposture persists after rhizotomy (Sherrington 1897, Denny-Brown 1980).

It should be stressed that gait equinus in our study was defined for a self-selected walking speed; a different distribution of obligate toe-strike among the 24 children might have been observed at different speeds of walking. This is an important consideration, since running by normal individuals results in a bilateral hemiplegic posture: arms flexed and adducted, with loss of swing, and legs adducted at the hip with a toe-strike. This suggests that a hemiplegic engram exists nor-

mally, to emerge physiologically when running, or pathologically after loss of normal cortical inhibition, *e.g.* stroke in childhood (Fig. 6). The often mild postural expression of hemiplegia in childhood can be revealed by getting the child to walk, run or perform the Fog test (Fog and Fog 1963, Gamstorp 1985)—*i.e.* the threshold for the hemiplegic posture varies between resting and active states. In contrast, the persistent hemiplegic posture seen after adult strokes is rare.

Gait equinus: an abnormal central pattern of walking?

The cerebral cortex normally activates muscles in the legs in a preset distal-to-proximal sequence (Nashner 1985). Leonard and colleagues (1991), in a recent kinematic and EMG study of the transition from supported to unsupported walking by normal and cerebral-palsied children, have shown that both groups have a common pattern of supported walking, which perists when children with cerebral palsy walk independently.

Hemiplegic side, hemiatrophy and cerebral plasticity

There was no relationship between gait equinus or passive equinus and the side of hemiplegia. Nor was limb atrophy more likely with lesions of one or other cerebral hemisphere. A study by Wiklund and Uvebrant (1991) could find no specific patterns of cerebral morphology associated with lower-limb growth or neurological abnormality in their 111 cases of hemiplegia: *i.e.* no support for the hypothesis of parietal wasting.

The lack of a lateral specificity for hemiatrophy in children may testify to the plasticity of the immature nervous system injured before the development of cerebral dominance. The varying degree of hemiatrophy, with muscle wasting and vasomotor instability, may be determined partly by a variable ipsilateral contribution from the undamaged hemisphere. Another possible mechanism is the branching of fast-conducting corticospinal pathways, providing bilateral presynaptic input to the motor neuron pool as a plastic response of the immature nervous system to injury (Farmer *et al.* 1990) and protecting the limb from atrophy.

Motor dexterity

There was no clear association between reduced fine-motor dexterity and gait equinus or passive equinus. This finding was expected, since fine-motor dexterity was universally reduced in all the affected limbs and could scarcely account for differences between one hemiplegia and another.

Passive equinus and peripheral variables

In contrast to gait equinus, there was a positive association between passive equinus (reduced compliance) and a shorter leg, reduced ankle-joint range, reduced extensibilité and increased resistance to passive stretch corresponding to a clinical impression of tonic spasticity.

Sharrard (1964) analysed the relationship between the degree of phasic spasticity—graded as mild, moderate or severe—and equinus deformity in 104 spastic limbs, but was unable to find any correlation between the two. Two-thirds of our hemiplegic children had evidence of reduced compliance. A more detailed analysis of our findings on muscle tone and reflex excitability is to follow, but we were unable to correlate resistance to passive stretch at rest with EMG activity, *i.e.* muscle contraction in the stretched muscles. These findings suggest a peripheral biomechanical transformation of muscle or changes in the plastic properties, rather than spasticity, as a cause of the resistance to passive movement at the ankle joint in childhood hemiplegia (see Fig. 4).

Implications for management of equinus in hemiplegia

The current evidence suggests that the principal causes of gait equinus are: (1) the expression of the hemiplegic posture to varying degrees; (2) a disturbance in central motor activation patterns; and (3) an abnormal phasic 'kick' produced by toe-strike, sustaining equinus during stance. Muscle power and muscle tone do not appear to contribute directly to gait equinus. The classical concepts of pathologically strong, spastic, shortened calf-muscles and weakened, elongated

APPENDIX TABLE A
Details of hemiplegic children

Case	*Age (yrs)*	*Sex* M	F	*Aetiology*[1] *Congenital*	*Perinatal*	*Acquired*	*Hemiside* R	L	*Seizures*	*Special educ.*	*Gait grade*[2] 1	2	3	4	*Muscle tone* *Phasic spasticity*	*Tonic spasticity*	*Hemi-dystonia*	*Normal tone*
1	9·2	—	+	Preterm			+	—	—	—	—	+	—	—	—	—	—	+
2	11·0	+	—	IUGR			+	—	+	+	—	—	—	+	+	+	—	—
3	11·5	+	—	Silent			+	—	—	—	—	+	—	—	—	—	—	+
4	11·7	+	—	IUGR			+	—	—	—	—	—	—	+	—	+	—	—
5	6·7	—	+		Asphyxia		+	—	—	—	—	—	—	+	—	—	—	+
6	9·3	—	+	Silent			+	—	—	—	+	—	—	—	+	+	—	—
7	11·4	+	—	Silent			—	+	—	—	—	+	—	—	—	—	—	+
8	10·2	—	+		Asphyxia		+	—	+	+	—	+	—	—	—	—	—	+
9	6·3	+	—	Silent			—	+	—	—	—	—	—	+	—	+	—	—
10	6·4	+	—	Silent			+	—	—	—	+	—	—	—	—	+	—	—
11	6·7	+	—	Silent			+	—	+	+	—	—	—	+	—	+	—	—
12	11·3	—	+	Preterm			+	—	—	—	—	+	—	—	+	+	—	—
13	11·8	+	—			Encephalitis	+	—	—	+	—	—	+	—	—	—	—	+
14	6·7	+	—	Silent			+	—	—	—	—	—	+	—	—	+	—	—
15	11·1	+	—	Silent			—	+	—	—	—	+	—	—	—	+	—	—
16	8·2	+	—	Preterm			+	—	—	—	—	—	—	+	+	+	—	—
17	6·2	+	—	Silent			+	—	—	—	—	—	—	+	+	+	—	—
18	12·0	+	—	IUGR			—	+	—	—	—	—	+	—	+	+	—	—
19	13·7	—	+			Rasmussen	—	+	+	+	—	—	—	+	+	—	—	—
20	12·9	+	—			Trauma	—	+	+	+	—	+	—	—	+	—	—	—
21	6·3	—	+			Post-op TOF	—	+	—	—	—	—	—	+	—	—	+	—
22	7·6	+	—		Asphyxia		—	+	—	—	—	—	+	—	+	+	—	—
23	12·2	—	+	Silent			—	+	—	—	+	—	—	—	—	—	+	—
24	17·7	+	—		Asphyxia		—	+	+	+	—	—	+	—	+	—	—	—
Total	9·9 (3·0)	16	8	16	4	4	14	10	5	7	3	7	5	9	10	14	2	6

[1]Silent = presumed silent infarct; IUGR = intra-uterine growth retardation; Encephalitis = post-measles; Rasmussen = encephalitis with intractable seizures requiring motor-strip topectomy age three, callosotomy age 10; Post-op TOF = cardiac by-pass surgery for total correction of tetralogy of Fallot.
[2]Gait score: 1 = heel-strike, 2 = plantar-strike, 3 = toe-heel strike, 4 = toe-strike.

APPENDIX, TABLE B

Goniometry at ankle for normal *vs.* affected side and right *vs.* left hemiplegia

Variable (N=23)	*Normal ankle Mean (SD)*	*Hemiplegic ankle Mean (SD)*	*t test*
Joint range	57·6 (6·1)	53·7 (6·1)	$p<0·03$

Variable (N=24)	*Right hemiplegia*			*Left hemiplegia*		
	Hemi ankle Mean (SD)	*Norm ankle Mean (SD)*	*t test*	*Norm ankle Mean (SD)*	*Hemi ankle Mean (SD)*	*t test*
Joint range*	52·5 (6·1)	58·9 (4·9)	$p<0·006$	63·3 (9·7)	55·6 (5·8)	NS
Joint range	52·5 (6·1)				55·6 (5·8)	NS
Joint range		58·9 (4·9)		63·3 (9·7)		NS
Resting equinus†	35·7 (11·1)	32·1 (7·3)	NS	14·6 (18·6)	16·2 (20·6)	NS
Resting equinus	35·7 (11·1)				16·2 (20·6)	$p<0·003$
Resting equinus		32·1 (7·3)		14·6 (18·6)		$p<0·003$

Full range of normal limb just significantly greater than that of affected limb. Resting equinus of normal and affected limbs in left hemiplegia is less than half that for normal and affected limbs in right hemiplegia.
*Total passive ankle-joint range. (Excluding case 19.)
†Angle of plantarflexion at rest: child supine. (Including case 19.)

anterior tibial muscles producing equinus is at variance with what is now known about the MVC about the ankle joint, since an equinus posture, if anything, favours dorsiflexor power. Limb dwarfing, reduced ankle-joint range and diminished extensibilité are not causes of gait equinus, but consequences of the same cerebral injury producing gait equinus. These peripheral changes are most likely to affect the efficiency of walking and running by reducing the speed and increasing the energy cost of such activities.

Splinting the ankle with an ankle-foot orthosis (AFO) corrects the central disturbances of a hemiplegic posture and an abnormal walking engram, and neutralises the 'kick' produced by the spinally-mediated phasic reflex excitability, which may reduce the energy cost of walking. The AFO and inhibitory casting operate by chronically *loading* the calf muscles. A tendo-Achilles tenotomy or a muscle recession procedure, in contrast to splinting with an AFO or casting, results in chronic *unloading* and weakening of already weakened calf muscles. Tenotomy completely neutralises all central, spinal and peripheral contributions to hindfoot equinus, since the Achilles tendon is the final common path for all these variables.

The two main treatments available for equinus have opposite physiological effects on the muscle.

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Authors' Appointments
*J-P. Lin, M.R.C.P., Edinburgh University Faculty of Medicine George Guthrie Research Fellow;
J. K. Brown, F.R.C.P., Consultant Paediatric Neurologist;
Department of Paediatric Neurology, Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF.

**Correspondence to first author.*

SUMMARY
A detailed clinical examination of 24 hemiplegic children at a mean age of 9·9 years showed that nine had an obligate toe-strike when walking at a self-selected speed and this was maintained throughout stance. None of the 24 children had a fixed equinus. No association could be found between this pattern of gait equinus and lower-limb atrophy, reduced ankle-joint range, muscle extensibilité, power of dorsiflexors and plantarflexors or actual muscle imbalance at the ankle joint. Gait equinus was independent of reduced compliance of the calf muscles, of a clinical diagnosis of tonic spasticity, of fine-motor dexterity of the toes, and of the side of the hemiplegia. Gait equinus cannot be explained merely in terms of a central paralytic foot-drop. A developmental model of equinus is advanced.

RÉSUMÉ

Les mécanismes périphériques et centraux du l'équin postérieur du pied chez l'enfant hémiplégique

Un examen clinique détaillé de 24 enfants hémiplégiques d'un âge moyen de 9·9 ans a montré que neuf d'entre eux avaient une contraction d'appui obligé des orteils durant une marche à vitesse contrôlée, qui se poursuivait tout au long du pas. Aucun des 24 enfants ne présentait un équin fixé. Aucune association n'a pu être trouvée entre cette allure de marche en équin et une atrophie du membre inférieur, une réduction de l'ouverture articulaire de la cheville, l'extensibilité musculaire, la force des fléchisseurs ou extenseurs dorsaux, ou encore le déséquilibre des forces musculaires effectivement présent à la cheville. L'équin était indépendant d'une réduction de compliance des muscles postérieurs de la jambe, d'un diagnostic clinique de spasticité tonique ou de la dextérité motrice des orteils du côté hémiplégique. L'équin de démarche ne peut être expliqué simplement en terme d'une chute paralytique du pied d'origine centrale. Un modèle de développement pour l'équin est proposé.

ZUSAMMENFASSUNG

Der peripheren und zentralen Mechanismen des Spitzfusses bei Kindern mit Hemiplegie

Eine detailierte klinische Untersuchung bei 24 Kindern im mittleren Alter von 9·9 Jahren zeigte, daß neun beim Laufen mit einer selbstgewählten Geschwindigkeit 'an obligate toe strike' hatten, der in der Haltung beibehalten wurde. Keines der 24 kinder hatte einen fixierten Klumpfuß. Es fand sich kein Zusammenhang zwischen diesem Gangmuster und einer Atrophie der unteren Extremität, einem verminderten Bewegungsradius des Fußlenkes, der Muskeldehnbarkeit, der Muskelkraft der Dorsi- und Plantarflexoren oder eines gestörten Muskelgleichgewichtes am Fußgelenk. Der Klumpfußgang war nicht abhängig von einer verminderten Ansprechbarkeit der Wadenmuskulatur, von der klinischen Diagnose einer tonischen Spastizität oder einer feinmotorischen Koordination der Zehen oder von der Seite der Hemiplegie. Der Klumpfußgang kann nicht ausschließlich mit einem zentral paralytischen Hängefuß erklärt werden. Ein Entwicklungsmodell des Klumpfusses wird vorgestellt.

RESUMEN

Los mecanismos periféricos y centrales del equinismo en la hemiplegia infantil

Un detallado examen clínico de 24 niños hemiplégicos de un promedio de edad de 9,9 años mostró que nueve apoyaban el dedo gordo del pie al andar a una velocidad auto-seleccionada y que esto se mantenia durante la bipedestación. Ninguno de los 24 niños tenía un equino fijado. No se pudo hallar ninguna asociación entre este patrón de marcha equina y una atrofia de la extremidad inferior, reducción del juego del tobillo, extensibilidad muscular, fuerza en los dorsiflexores y plantaflexores o desequilibrio muscular a nivel del tobillo. La marcha equina era independiente de una complianza reducida de los músculos de l pantorrilla o de un diagnóstico clínico de espasticadad tónica, de una destreza motora fina de los dedos de los pies o del lado de la hemiplegia. Le marcha equina no puede ser explicada meramente en términos de un pie caído por parálisis central. Se propone un modelo de equinismo basado en el desarrollo.

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ASSESSMENT OF SPASTICITY IN HEMIPLEGIC CEREBRAL PALSY I: PROXIMAL LOWER-LIMB REFLEX EXCITABILITY

Jean-Pierre Lin
J. Keith Brown
Ruth Brotherstone

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The study of spasticity as a sign of disordered motor control has been in and out of fashion as an object of clinical interest and physiological study for more than a century. The advent of intrathecal baclofen, however, and the recent rehabilitation of Forster's rhizotomy in the form of selective posterior rhizotomy, have made it necesary to review the basic assumptions about 'the spasticities'. A clear definition of spasticity is essential to any argument about the benefits of one treatment over another (Landau 1974). The functional significance of spasticity as the result of damage to the growing and developing brain continues to be controversial. The patterns of disturbed tone arising from damage to the adult brain may differ from those of the child with cerebral palsy: indeed, recent evidence from work with children suggests that new pathways are laid (or their regression offset) by early injury to the motor system (Carr *et al.* 1991, Cohen *et al.* 1991, Farmer *et al.* 1991).

Muscle tone is defined as the resistance felt when a limb is passively rotated about a joint. The factors contributing to this resistance can be divided into (1) non-electrical biomechanical (plastic and visco-elastic) resistance, and (2) electrical contraction, whether mediated by a central or peripheral reflex, or by voluntary or involuntary activation (Table I)

There have been many studies on the nature of reflexes and reflex excitability over the last 30 years. The most commonly accepted definition of spasticity is that of Lance (1980): 'In both cerebral and spinal spasticity, the stretch reflex responses obtainable from extensor and flexor muscle groups of the upper and lower limbs increase approximately linearly with increase in the velocity of stretch. The reflex component of the increased tone may therefore be measured in terms of the threshold velocity required to evoke reflex activity and the slope of the EMG–velocity relationship'. It follows that the velocity threshold of stretch necessary to elicit a reflex is reduced, and the amplitude of a reflex response to a given velocity of stretch is increased in spasticity, compared with the normal.

A wide variability of responses is said to be the hallmark of normal reflex excitability, whereas the individual with spasticity exhibits a stereotyped exaggeration of reflex activity which is only occasionally encountered in normal individuals (Neilson and Lance 1978, Rack *et al.* 1984).

The use of step-torque averaging techniques (equivalent to a ramp stretch manoeuvre) have demonstrated that the upper latency of a true polysynaptic reflex response occurs in under 90ms when

actively contracting wrist flexors are perturbed (Lee and Tatton 1978), or 150ms for the triceps surae muscle (Berardelli *et al.* 1982). This methodology sets a latency limit on what may be regarded as stretch reflex activity: according to this definition, any EMG activity occurring outside 90ms for the wrist, or 150ms for the calf, is not a stretch reflex.

The study of reflex behaviour requires knowledge of the starting muscle length, which corresponds to the initial joint angle, the amplitude and velocity of stretch. The state of the muscle before stretch (whether resting or active) and the method used to stretch the muscle should be specified. By combining a sinusoidal with a ramp technique we believe that most aspects of monosynaptic reflex activity can be assessed. This paper presents a study of proximal lower-limb reflex excitability and is followed by a detailed study of calf stretch reflexes in childhood hemiplegia.

TABLE I
Mechanisms resisting passive muscle stretch

Biomechanical mechanisms resisting stretch
Elasticity: length-dependent
Viscosity: velocity-dependent
Inertia: acceleration-dependent
Plasticity: time-dependent (creep, stress-relaxation, thixotropy)
Contracture: of muscle/tendon/joint ligaments
Neural mechanisms resisting stretch
Reflex EMG: abolished by rhizotomy
Spasticity: velocity-dependent reflex contraction mediated by muscle spindles due to loss of presynaptic inhibition
Central EMG: unaffected by rhizotomy
Dystonia: fluctuations of tone dependent on position in space and non-specific afferent input and abnormal body-contact responses
Posture: hemiplegic posture
Rigidity: hypokinetic increased resistance
Tension: failure to relax
Gegenhalten: load-seeking behaviour?
Voluntary contraction: resist = increased tone, assist = reduced tone

Patients

A group of 13 children with congenital hemiplegia aged 6·2 to 12·2 (mean 9·4) years, described in a previous clinical study (Lin and Brown 1992), underwent evaluation of proximal lower-limb reflex excitability under electromyographic control. All had congenital hemiplegia. Perinatal details include preterm birth in two cases, intra-uterine growth retardation in two cases, and birth asphyxia in a further two cases. The remaining seven cases were born at term without known antenatal or perinatal difficulty. Further details of the patient population are given in the Appendix, including a clinical assessment of patterns of walking and muscle tone.

Method

The children lay comfortably prone on a couch watching a video of their own choice in a warm, quiet room in the presence of their parents. Their feet were off the couch, allowing a full knee-joint range from full extension to full flexion. The normal and affected limbs were compared. It took approximately one hour to investigate the thigh muscles of both legs for each child. Surface EMG was assessed with the child at rest. Disposable surface electrodes (Medicotest 'Blue Sensor') with a diameter of 5mm were arranged in a bipolar array with an inter-electrode distance of approximately 6 to 10cm, depending on the size of the child, along the long axis of the muscle, with the active electrode proximal to the reference. The earth electrode was placed either on the same or opposite limb. The skin was cleansed with an abrasive cream before electrode placement. The hamstrings and quadriceps muscles were studied as agonist–antagonist pairs. Electrodes were placed over the middle anterior and posterior thigh overlying each muscle. Electrode jelly to improve contact was used to supplement that provided on the disposable electrode. A piece of adhesive tape secured the position of the electrodes to the skin overlying the muscle of interest. The EMG signal was acquired and displayed on an eight-channel SLE 800 EEG machine with a bandwidth of 0·5 to 150Hz using a paper speed of 1·5cm/s and display gain of 10μV/mm.

Joint angles were measured using a flexible twin-axis Penny and Giles goniometer with a $\pm$5V output reduced to $\pm$500μV by a variable resistor before connection with the SLE recorder. The quadriceps and hamstring muscles were

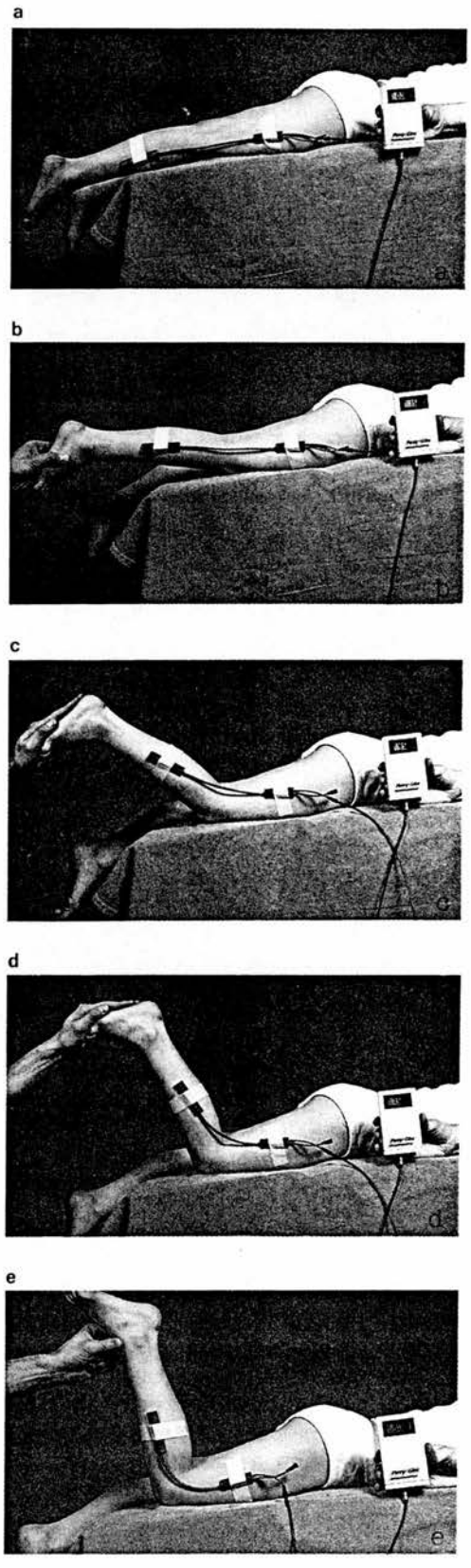

Fig. 1. Six-year-old boy with a congenital right hemiparesis lies prone on couch with a Penny and Giles twin-axis flexible goniometer attached to lateral knee joint, measuring angular displacement. (a) 0°, knee fully extended at rest; (b) 15°; (c) 45°; (d) 75°; and (e) 90°, knee flexion. Sinusoidal stretches of ±15° were imposed at centres of oscillation of 15°, 45° and 75°. Ramp stretches were performed from 0° to 30°, 30° to 60° and 60° to 90° for quadriceps ramp stretches and in reverse direction for hamstring ramp stretches. As knee is flexed, quadriceps lengthen and hamstrings shorten, and vice versa.

tested over a 90° range, 0° representing full knee extension with the goniometer taped across the lateral aspect of the knee joint. Figure 1 shows the principal joint positions of 15°, 45° and 75° about which the muscles were stretched over approximately a 30° arc. Each goniometer trace was sandwiched between the EMG signals of the muscles to allow EMG interpretation in relation to displacement amplitude and angular velocity of ramp stretch or frequency of sinusoidal oscillation. The goniometer output was calibrated with a plastic goniometer which was used to give a calibration signal to the SLE recorder of 40° peak-to-peak and allowed cross-reference with the goniometer's own instantaneous liquid crystal display. The signal error was ±1 to 2°.

Stretch protocol

RAMP STRETCH

Each muscle was subjected to sudden ramp stretches at amplitudes of about 30° displacement at varying angular velocities of stretch. At the knee, the ramp flexions applied to the quadriceps were approximately 0 to 30°, 30 to 60° and 60 to 90°. For the hamstrings, ramp extensions from 90 to 60°, 60 to 30° and 30 to 0° were applied.

SINUSOIDAL STRETCH

Each muscle underwent manual sinusoidal stretch at rest according to the method of Burke *et al.* (1971), at amplitudes of ±15° about three centres of oscillation: 15°, 45° and 75° of knee flexion (Figs. 1b to d). Reflex excitability is known to vary with initial muscle length. The centre of oscillation determines the starting length of the

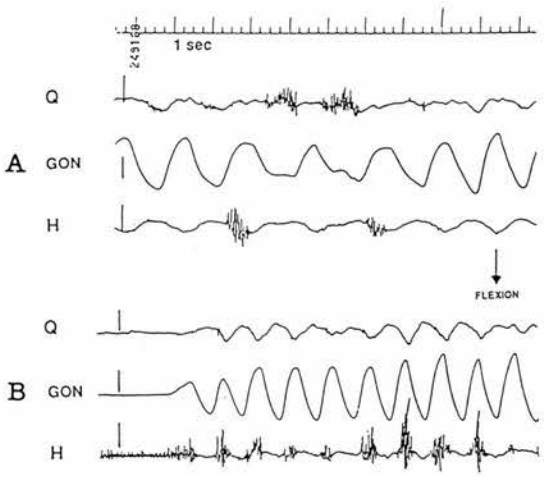

Fig. 2. Twelve-year-old boy (case 18) with congenital left hemiplegia. (A) Voluntary resist movements distort sinusoidal motion applied by examiner's hand: note actively contracting quadriceps (Q) and hamstrings (H). Goniometer (GON) waveform is particularly distorted in flexion. (B) Initially active hamstring (H) muscles lead to an augmented EMG each time they are stretched (knee extension), and to a silent period with knee flexion (flexion in direction of arrow*). Calibration bars: EMG 100μV, goniometer 20°; centre of oscillation 45°.*

muscles and allows comparison of reflexes at equivalent muscle lengths between subjects. For any pair of agonist–antagonist muscles, the shortest position of the one is the longest position of the other. The frequency of sinusoidal oscillation varied from 0·5 to 3·5Hz at the knee. Conformity to a sinusoidal waveform was maintained according to the goniometer output trace. The torque required to displace the limbs at any given amplitude and frequency was applied manually by the examiner, but not measured. Since the inertia of the limb varies with the square of the frequency, only relatively low frequencies of stretch are possible with manual stretch.

Wartenberg's pendulum test

Knee jerks were elicited with the children dangling their legs over the edge of the couch and monitored with goniometry and EMG. The maximum duration of the damped oscillation in seconds and maximum amplitude of swing in degrees following a tap to the patella tendon were measured. Traces which showed evidence of restlessness before or after taps were discarded.

EMG analysis

All the tests were performed by the same examiner (J.-P.L.), and electrode placement and operation of the SLE recorder were performed under the supervision of the same technician (R.B.). All the traces of EMG activity were analysed according to the following criteria:

(1) EMG silence was a prerequisite before either ramp or sinusoidal stretching. Sequences containing EMG activity before stretch were discarded as representing inadequate relaxation or involuntary activity.

(2) Voluntary or involuntary co-contractions during sinusoidal stretch abolish or distort displacement and show augmented tonic EMG at maximum stretch and an EMG silent period (Fig. 2) when the muscle is unloaded (Angel 1973, Struppler *et al.* 1973).

(3) During sinusoidal stretch, a triphasic agonist–antagonist EMG response was interpreted as characteristic of voluntary alternating flexion and extension movements. This usually occurred at low-frequency stretch cycles, when subjects could anticipate the direction of movement.

(4) Reflex excitability was measured in terms of the velocity or frequency threshold and the initial muscle length as defined by the centre of oscillation at which a reflex was elicited.

(5) The term 'non-paretic muscles' refers to the unaffected limb of a hemiparetic child. Non-paretic and hemiparetic reflex thresholds were compared at equivalent initial joint positions.

Results

Hamstrings and quadriceps muscles were subjected to rapid ramp stretch from rest in 12 cases. Results are expressed in terms of the minimum angular velocity threshold required to elicit a reflex EMG

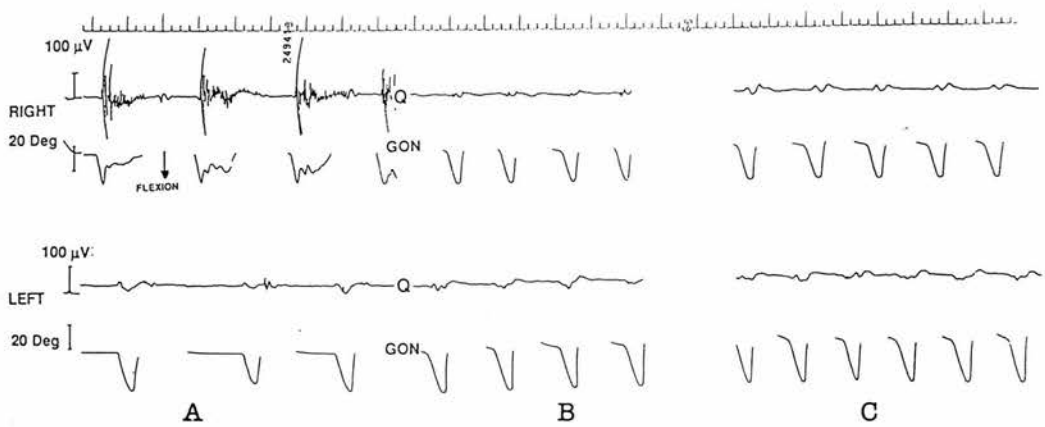

*Fig. 3. Reflex velocity threshold following phasic ramp stretch of normal (*LEFT*) and hemiparetic (*RIGHT*) quadriceps. 10-year-old girl with congenital right hemiplegia. (A) 0° to 30°; (B) 30 to 60°; (C) 60° to 90° of rapid knee flexion. Note quadriceps (Q) EMG in response to phasic stretch of hemiparetic muscle at its shortest length close to full knee extension; no response of affected Q at other lengths. Normal Q is unresponsive at all lengths.* GON = *goniometer. Calibration bars: EMG 100μV, goniometer 20°, 1s divisions.*

TABLE II

Comparison of the mean reflex velocity thresholds for the non-paretic and hemiparetic quadriceps (Q) and hamstring (H) muscles during rapid (phasic) ramp stretch with increasing muscle length in childhood hemiplegia

Muscle		*Cases (N)*	*Ramp stretch (knee flexion)*	*Reflex threshold reached*	*Velocity ramp stretch (°/s)*		*Confidence interval*		*Paired t test, (p value)*
					Mean	*(SD)*	*Lower (95%)*	*Upper (95%)*	
Non-paretic	Q	8	0–30°	+	131·3	(33·72)	103·1	159·5	
Non-paretic	Q	5	30–60°	+	146·0	(31·29)	107·1	184·9	NS
Non-paretic	Q	2	60–90°	+	208·5	(61·51)	−344·2	761·2*	
Hemiparetic	Q	10	0–30°	+	124·7	(28·12)	104·5	144·8	
Hemiparetic	Q	6	30–60°	+	148·3	(34·79)	111·8	184·8	<0·01
Hemiparetic	Q	5	60–90°	+	136·6	(33·99)	94·3	178·8	NS
Non-paretic	H	8	30–0°	+	112·8	(22·1)	94·3	131·4	
Non-paretic	H	7	60–30°	+	131·5	(37·1)	97·1	165·9	<0·01
Non-paretic	H	4	90–60°	+	119·0	(40·24)	54·96	183·0	NS
Hemiparetic	H	8	30–0°	+	101·1	(22·5)	82·2	119·9	
Hemiparetic	H	7	60–30°	+	130·0	(36·34)	96·3	163·6	<0·01
Hemiparetic	H	7	90–60°	+	156·7	(55·6)	105·2	208·2	<0·02

*Insufficient numbers for paired *t* test.

response. Figure 3 shows the EMG and goniometer traces from a 10-year-old girl with a congenital right hemiplegia for non-paretic and hemiparetic quadriceps ramp stretches at three successive knee-joint positions.

Quadriceps reflex velocity threshold with muscle length

Table II gives the results of the mean reflex velocity thresholds for the non-paretic and hemiparetic quadriceps muscles in each of the children according to initial muscle length. As the quadriceps is lengthened, the reflex velocity threshold increases. This reduced velocity sensitivity with increasing muscle length achieved statistical significance for the hemiparetic quadriceps ($p < 0{\cdot}01$), but the non-paretic quadriceps did not show a similar significant length-dependence. No differences in reflex velocity thresholds

TABLE III

Mean reflex velocity thresholds and maximum velocities for which no reflex threshold was reached following rapid (phasic) ramp stretch of quadriceps (Q) and hamstring (H) muscles in childhood hemiplegia

Muscle		*Cases (N)*	*Ramp stretch (knee flexion)*	*Reflex threshold reached*	*Velocity ramp stretch (°/s)*		*Confidence interval*		*Paired t test, (p value)*
					Mean	*(SD)*	*Lower (95%)*	*Upper (95%)*	
Non-paretic	Q	8	0–30°	+	131·3	(33·72)	103·1	159·5	NS
Hemiparetic	Q	10	0–30°	+	124·7	(28·12)	104·5	144·8	
Non-paretic	Q	4	0–30°	–	116·7	(25·34)	76·42	157·0	*
Hemiparetic	Q	0	0–30°	–	—	—	—	—	
Non-paretic	Q	5	30–60°	+	146·0	(31·29)	107·1	184·9	NS
Hemiparetic	Q	6	30–60°	+	148·3	(34·79)	111·8	184·8	
Non-paretic	Q	7	30–60°	–	135·0	(37·72)	100·1	169·8	NS
Hemiparetic	Q	4	30–60°	–	144·4	(28·89)	98·5	190·4	
Non-paretic	Q	2	60–90°	+	208·5	(61·51)	−344·2	761·2	*
Hemiparetic	Q	5	60–90°	+	136·6	(33·99)	94·3	178·8	
Non-paretic	Q	9	60–90°	–	138·2	(57·16)	94·2	182·1	NS
Hemiparetic	Q	5	60–90°	–	187·2	(73·18)	96·3	278·0	
Non-paretic	H	8	30–0°	+	112·8	(22·1)	94·3	131·4	NS
Hemiparetic	H	8	30–0°	+	101·1	(22·5)	82·2	119·9	
Non-paretic	H	0	30–0°	–	—	—	—	—	*
Hemiparetic	H	0	30–0°	–	—	—	—	—	
Non-paretic	H	7	60–30°	+	131·5	(37·1)	97·1	165·9	NS
Hemiparetic	H	7	60–30°	+	130·0	(36·34)	96·3	163·6	
Non-paretic	H	2	60–30°	–	167·5	(34·64)	−143·8	478·8	*
Hemiparetic	H	1	60–30°	–	143·0	—	—	—	
Non-paretic	H	4	90–60°	+	119·0	(40·24)	54·96	183·0	NS
Hemiparetic	H	7	90–60°	+	156·7	(55·6)	105·2	208·2	
Non-paretic	H	5	90–60°	–	168·8	(53·93)	101·8	235·7	*
Hemiparetic	H	1	90–60°	–	180·0	—	—	—	

*Insufficient numbers for paired *t* test.

between non-paretic and hemiparetic quadriceps muscles could be established at comparable knee-joint positions (Table III).

Hamstring reflex velocity threshold with muscle length

The hamstring reflex velocity threshold diminishes with increasing muscle length—*i.e.* as the knee position moves towards full extension, the hamstrings become more excitable—and this was moderately significant for the non-paretic ($p<0·01$) and hemiparetic ($p<0·01$) hamstrings (Table II). Non-paretic and hemiparetic hamstrings appeared to have a similar reflex velocity threshold at comparable muscle lengths (Table III).

Variations in reflex velocity threshold with initial muscle length

Table III indicates those non-paretic and hemiparetic hamstrings and quadriceps muscles for which no reflex velocity threshold could be elicited. At each range of stretch, these muscles were subjected to stretch velocities that were similar to or higher than those in muscles for which a reflex response was obtained. The proportion of muscles for which it is not possible to elicit a reflex velocity threshold increases with increasing quadriceps length and hamstring shortening. Overall, the quadriceps and hamstring muscles were most velocity-sensitive—*i.e.* were most reactive—close to the position of full knee extension, but non-paretic and

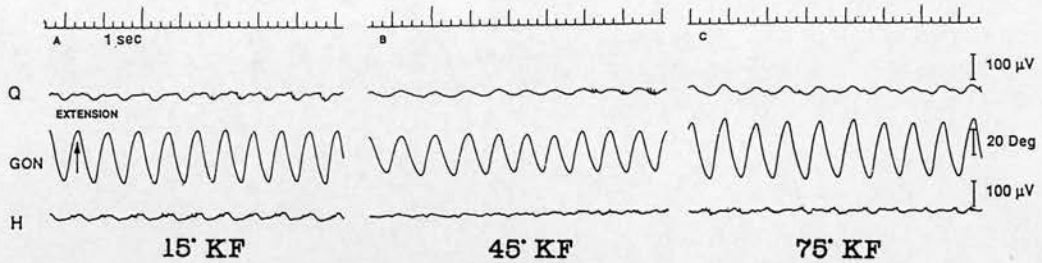

Fig. 4. Sinusoidal stretch of hemiparetic quadriceps (Q) and hamstrings (H) at three centres of oscillation (CO) in 10-year-old girl with congenital right hemiplegia. GON = *goniometer,* KF = *knee flexion.*

CO		*Reflex frequency threshold*	
		Q	*H*
A	15°	1·3Hz	1·3Hz
B	45°	1·3Hz	1·3Hz
C	75°	*	*

*No reflex threshold reached at 75° *CO* frequency of oscillation 1·3Hz shown.

TABLE IV

Comparison of the mean reflex frequency thresholds for the non-paretic and hemiparetic quadriceps (Q) and hamstring (H) muscles during sinusoidal oscillation at increasing muscle lengths in childhood hemiplegia

Muscle		*Cases (N)*	*Centre of oscillation (degrees of knee flexion)*	*Reflex threshold reached*	*Frequency of oscillation (Hz)*		*Confidence interval*		*Paired t test, (p value)*
					Mean	*(SD)*	*Lower (95%)*	*Upper (95%)*	
Non-paretic	Q	8	15°	+	2·0	(0·59)	1·5	2·5	
Non-paretic	Q	6	45°	+	1·7	(0·37)	1·3	2·1	NS
Non-paretic	Q	0	75°	+	—	—	—	—	*
Hemiparetic	Q	6	15°	+	2·1	(0·39)	1·7	2·5	
Hemiparetic	Q	8	45°	+	2·0	(0·62)	1·49	2·5	NS
Hemiparetic	Q	1	75°	+	1·2	—	—	—	*
Non-paretic	H	8	15°	+	1·8	(0·54)	1·3	2·2	
Non-paretic	H	6	45°	+	1·7	(0·37)	1·3	2·1	NS
Non-paretic	H	3	75°	+	2·0	(0·83)	0·0	4·1	NS
Hemiparetic	H	9	15°	+	1·3	(0·34)	1·1	1·6	
Hemiparetic	H	8	45°	+	2·0	(0·6)	1·4	2·5	<0·05
Hemiparetic	H	5	75°	+	1·8	(0·72)	0·9	2·7	NS

*Insufficient numbers for paired *t* test.

hemiparetic muscle groups had similar reflex velocity thresholds (Table III).

Sinusoidal stretch

Sinusoidal stretching of quadriceps and hamstring muscles was performed in 13 children. Results are expressed as the minimum frequency of sinusoidal oscillation required to elicit a reflex EMG response in the quadriceps and hamstrings: the reflex frequency threshold. The centre of oscillation determines the initial muscle length (Figs. 1b to d). The amplitude of sinusoidal displacement about any given centre of oscillation was 20 to 50°, keeping as close to 30° as possible. Figure 4 shows the EMG and goniometer output for the hemiparetic quadriceps of the same 10-year-old girl with a congenital right hemiplegia as in Figure 3. A reflex frequency threshold to sinusoidal stretch of 1·3Hz was reached when the quadriceps muscle was stretched about centres of oscillation of 15 and 45°

TABLE V

Mean reflex frequency thresholds and maximum frequencies of sinusoidal oscillation for which no reflex was elicited in non-paretic and hemiparetic quadriceps (Q) and hamstring (H) muscles in childhood hemiplegia

Muscle		*Cases (N)*	*Centre of oscillation (degrees of knee flexion)*	*Reflex threshold reached*	*Frequency of oscillation (Hz)*		*Confidence interval*		*Paired t test, (p value)*
					Mean	*(SD)*	*Lower (95%)*	*Upper (95%)*	
Non-paretic	Q	8	15°	+	2·0	(0·59)	1·5	2·5	NS
Hemiparetic	Q	6	15°	+	2·1	(0·39)	1·7	2·5	
Non-paretic	Q	5	15°	–	2·1	(0·75)	1·1	3·0	
Hemiparetic	Q	7	15°	–	1·2	(0·29)	0·9	1·5	NS
Non-paretic	Q	6	45°	+	1·7	(0·37)	1·3	2·1	NS
Hemiparetic	Q	8	45°	+	2·0	(0·62)	1·49	2·5	
Non-paretic	Q	7	45°	–	2·1	(0·79)	1·5	2·8	NS
Hemiparetic	Q	4	45°	–	2·0	(0·05)	1·9	2·1	
Non-paretic	Q	0	75°	+	—	—	—	—	*
Hemiparetic	Q	1	75°	+	1·2	—	—	—	
Non-paretic	Q	13	75°	–	2·0	(0·44)	1·7	2·3	NS
Hemiparetic	Q	11	75°	–	2·2	(0·55)	1·8	2·5	
Non-paretic	H	8	15°	+	1·8	(0·54)	1·3	2·2	<0·05
Hemiparetic	H	9	15°	+	1·3	(0·34)	1·1	1·6	
Non-paretic	H	5	15°	–	2·2	(0·97)	1·0	3·4	NS
Hemiparetic	H	4	15°	–	1·5	(0·25)	1·1	1·9	
Non-paretic	H	6	45°	+	1·7	(0·37)	1·3	2·1	NS
Hemiparetic	H	8	45°	+	2·0	(0·6)	1·4	2·5	
Non-paretic	H	7	45°	–	2·1	(0·72)	1·5	2·8	NS
Hemiparetic	H	4	45°	–	2·0	(0·05)	1·9	2·1	
Non-paretic	H	3	75°	+	2·0	(0·83)	0·0	4·1	NS
Hemiparetic	H	5	75°	+	1·8	(0·72)	0·9	2·7	
Non-paretic	H	10	75°	–	2·0	(0·35)	1·7	2·3	NS
Hemiparetic	H	7	75°	–	2·1	(0·28)	1·8	2·4	

*Insufficient numbers for paired *t* test.

of knee flexion, but the same frequency failed to elicit a reflex when the muscle was stretched at 75° of flexion.

Quadriceps reflex frequency threshold and muscle length

The mean reflex frequency threshold for non-paretic and hemiparetic quadriceps muscles in 13 children is shown in Table IV at three knee-joint positions (centres of oscillation): 15, 45 and 75° of knee flexion. As for ramp stretches, the number of cases in whom a reflex frequency threshold could be established declined as the centre of oscillation approached 90° of flexion. There were no differences in reflex frequency threshold between non-paretic and hemiparetic quadriceps at any given muscle length (Table V).

Hamstring reflex frequency threshold and muscle length

Similar mean sinusoidal stretch frequencies elicited a hamstring reflex at the three centres of oscillation (Table IV). There was a weakly significant lowering of the hamstring reflex frequency threshold on paired *t* testing for hemiparetic compared with non-paretic hamstrings ($p<0·05$) close to full knee extension (Table V), but this difference was not seen at shorter hamstring positions.

Variation in reflex frequency threshold with muscle length

For sinusoidal oscillation, as with phasic ramp stretching, the number of cases in which it was possible to establish a reflex frequency threshold in quadriceps and hamstrings declined with increasing knee

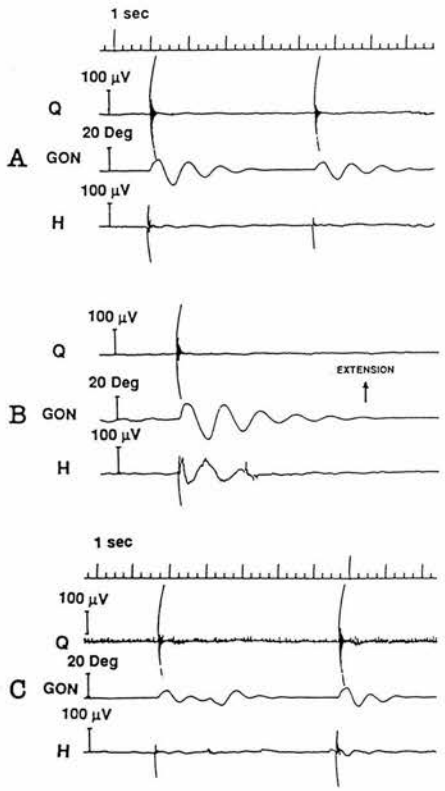

Fig. 5. (A) Hemiparetic limb. (B) Non-paretic limb. Note that quadriceps (Q) and hamstring (H) EMGs are silent before knee jerk in (A) and (B) and ensuing pendular movements are smooth and eventually dampen. Continuous EMG activity of Q before tendon jerk in (C) distorts pendular swing of knee (GON), and such traces are discarded. Maximum amplitude of swing and duration of damped oscillation were measured in seven cases (see Table VI), but no differences between non-paretic and hemiparetic muscles were found.

flexion. The maximum frequencies of sinusoidal stretch to which these muscles were exposed were similar to those at which a threshold response was obtained (Table V).

Comparison of ramp vs. *sinusoidal stretching*

Although rapid ramp stretches showed a closer relationship between reflex velocity threshold and initial muscle length, no differences in reflex velocity threshold between non-paretic and hemiparetic limbs were apparent at any given muscle length. Sinusoidal stretching was able to demonstrate a weakly significant lowering of the hamstring reflex frequency threshold in the hemiparetic compared with non-paretic hamstrings at a position close to full knee extension only, but no such difference could be found for the quadriceps.

Wartenberg's pendulum test

Knee jerks were elicited in 12 cases (Fig. 5) and the means and 99 per cent confidence intervals for the duration of damped oscillation and maximum amplitude of swing are shown in Table VI, but there were no significant differences between sides. In five cases, one or other limb was restless during the test so that only seven of 12 paired comparisons between non-paretic and hemiparetic legs were possible.

Discussion

Our findings demonstrate little reflex excitability of the thigh muscles in hemiplegic cerebral palsy using either sudden ramp or continuous, rhythmic sinusoidal methods of stretch. The hamstrings showed greater velocity sensitivity than the quadriceps when subjected to stretch at different initial muscle lengths. However, with the exception of the hemiparetic hamstrings close to full knee extension, hemiplegic muscles were no more excitable than the non-paretic muscles when compared length for length. An assessment of traditional knee jerks using Wartenberg's pendulum test failed to demonstrate differences in excitability to a supra-maximal stimulus between sides. This implies a similar processing by the spinal cord of afferent input from the muscle spindles: for instance, a similar degree of presynaptic inhibition.

There may be several explanations for these results, each of which is examined below.

(1) Hemiparetic muscles are more excitable, but the method of reflex EMG detection was too insensitive. Since the skin and underlying tissues act as low-frequency (high-pass) filters for the EMG signal (Basmajian and de Luca 1985), we do not believe that much EMG activity was 'missed' using the SLE 800 recorder. There is a long list of disadvantages with needle or wire electrodes compared with

TABLE VI

Wartenburg's pendulum test at the knee: comparison of duration of damped oscillation and maximum amplitude of swing for non-paretic and hemiparetic legs following clinical knee jerks in childhood hemiplegia

Knee jerk	*Non-paretic (N = 10)*	*Hemiparetic (N = 9)*	*Paired t test (N = 7)*
Duration of damped oscillations (s)			
Mean (SD)	4·22 (1·36)	3·92 (1·14)	NS
99% Lower CI	3·11	2·94	
99% Upper CI	5·32	4·91	
Amplitude of maximum swing (°)			
Mean (SD)	21·62 (16·77)	17·80 (8·72)	NS
99% Lower CI	7·95	10·36	
99% Upper CI	35·28	25·34	

Tendon taps were applied to patella tendons of children seated with legs dangling freely at rest. All 12 cases were tested. Traces with evidence of restlessness before and after taps were discarded. A paired comparison of seven cases showed no statistical difference in duration of damped oscillation or amplitude of swing between non-paretic and hemiparetic legs.

surface EMG, including pain and discomfort, noisy signals during imposed movements and the fact that only muscles within 100μm distance of the electrode are recorded. The sensitivity of 10μV/mm appears reasonable for detecting low-amplitude signals. Very small signals, recorded during a study of Wartenberg's pendulum test in the same children, were sufficient to alter the profile of damped oscillations at the knee (unpublished observation).

(2) The method of stretching the muscles may have failed to elicit a reflex response, but both ramp stretching and sinusoidal oscillation are extensions of the bedside clinical method for assessing tone. In this study, the position of the knee and the electrical changes of the muscle were recorded without discomfort to the child, and the approach to stretching was standardised. It is inevitable that small variations occurred from muscle to muscle or subject to subject which might have been eliminated by mechanical stretching techniques using a torque motor, but the advantages of a clinical method capable of being used in any setting would have been lost.

(3) It could be claimed that the principle of studying muscles from rest is unphysiological, whereas assessing partially active muscles might have shown more reflex excitability. Many studies relying on minimally contracting muscles (usually 10 to 20 per cent of maximum voluntary contraction) have demonstrated how differently the motor system can behave, depending on the extent of muscle preactivation. Cutaneomuscular reflexes (Evans *et al.* 1991), long-latency stretch reflexes (Berardelli *et al.* 1982, 1983) and magnetic brain stimulation (O'Sullivan *et al.* 1991) all require active muscles. However, the difficulty lies in standardising the level of baseline muscle activity, which requires a method of signal averaging over many cycles. At the outset of this study it was anticipated that the resistance to slow stretch would be a combination of reflex contraction and biomechanical resistance. Provided the children were completely relaxed, there was little reflex EMG whatever the method of muscle stretch. It was easy to disregard sequences in which continuous EMG activity preceded stretch. Voluntary contamination of an EMG sequence could usually be distinguished from true reflex activity, if during a sinusoidal stretch sequence the EMG was originally silent and became briefly active (see Fig. 2a). Actively contracting muscles from either the non-paretic or hemiparetic limbs all showed an augmented reflex response to stretch (see Fig. 2b), with a corresponding unloading response producing a muscle 'silent period' (Angel 1973, Struppler

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et al. 1973) which became undetectable when muscle preactivation ceased.
(4) Burke *et al.* (1971) and Rack *et al.* (1984) reported fatigue in their studies of sinusoidal stretch in the thigh and calf. From our records, we were unable to determine any pattern consistent with fatigue.
(5) It has been demonstrated that the stretch-evoked cortical potential diminishes with an increasing frequency of presentation of the stretch stimulus of wrist extensors (Rothwell *et al.* 1987). This results in a long-latency EMG response of smaller amplitude as the stimulus frequency increases from 0·1 to 1Hz. By definition, monosynaptic short-latency responses are unaffected. Our study did not attempt to evaluate long-latency responses, so it is unlikely that our results were affected by problems of central motor habituation.
(6) There is always the possibility of a larger study producing statistically significant results that are clinically irrelevant. This study involved repeated measurements on four muscles in each of 13 subjects at three different muscle lengths, using two distinct methods of stretch. This is equivalent to about 156 non-paretic muscles being compared with 156 hemiparetic muscles.

All the possible methodological pitfalls mentioned above would still need to explain why the hemiparetic and non-paretic EMG responses to stretch were essentially similar.

Biomechanical vs. *reflex resistance*

This relatively electrically silent resistance to stretch at rest suggests that the resistance felt must be biomechanical in nature, even when the knee joint is close to full extension and the quadriceps and hamstrings are at their most excitable. This implies that the increased resistance, or 'stiffness', may be due to peripheral muscular transformation of either the contractile elements of muscle or adjacent non-contractile elements. Evidence for a lack of reflex activity in resting 'spastic' muscles was confirmed by Foley (1961). He demonstrated that the velocity-dependent hypertonus in resting normal and spastic adults and children with spastic diplegia was independent of reflex EMG activity. He showed very clearly the phenomenon of 'stress relaxation', which is the gradual reduction in torque required to maintain a given muscle length over time. He also showed that physical therapy involving limb and trunk stretching could significantly reduce muscle tone, whereas complete peripheral nerve blocks had no effect.

Why should children be different from adults?

There are several possible reasons why the effects of central motor injury in adults differ from those which arise from congenital injuries or those sustained in early infancy. Herman (1970) showed differences in the reflex and the visco-elastic properties of muscle in relation to duration and severity of the stroke symptoms. Similarly, Thilmann *et al.* (1991) demonstrated that maximum EMG activity occurred between the first and second month following a stroke in the biceps muscles of 19 hemiparetic adults in their study, and that it declined thereafter during the first year. This evidence suggests that reflex excitability diminishes with time, or 'burns out', and this may equally apply to children, though in most cases the clinical patterns of reflexes were never particularly brisk. Evidence is emerging that the growing and developing brain responds to damage in some cases by the fast-conducting cortical motor units of one hemisphere supplying bilateral lower motor neuron pools (Farmer *et al.* 1991) and sometimes producing mirror movements.

In an adult who had undergone a left hemispherectomy for intractable seizures secondary to a congenital porencephalic cyst, Cohen *et al.* (1991) demonstrated activation of the biceps and deltoid of the hemiparetic arm when the surviving cortex was magnetically stimulated 2cm anterolateral to the point of activation of the non-paretic side. Voluntary movements of the affected limb produced an increase in cerebral blood flow in an area 1·5cm anterior to that produced by movement of the non-paretic arm and also produced a pre-movement potential in the surviving ipsilateral hemisphere. This cerebral plasticity appears to be confined to motor reorganisation, since

no sensory evoked potentials to median nerve stimulation could be elicited from the hemiparetic limb. This motor reorganisation may prevent the loss of presynaptic inhibition which results from damage to supraspinal structures in adults. Evidence supporting plastic reorganisation of the brain comes from the observation that clinical signs, function and disability appear to bear little relation to the size and distribution of the lesion on CT imaging (Wiklund and Uvebrant 1991) in hemiplegic cerebral palsy. Another possible palliating factor stems from the fact that the principal cause of congenital hemiplegia appears to be placental embolism and the intrauterine environment may provide support in maintaining homeostasis even in the presence of a fetal stroke, offsetting secondary hypoxic–ischaemic injuries.

Of the 18 cases studied by Burke *et al.* 1971, only one case had spasticity of cerebral origin attributed to a parasagittal meningioma, and in five of the 18 cases it was not possible to elicit a sinusoidal reflex threshold. These adult patients appeared to have much lower reflex-frequency thresholds (of the order of 0·5Hz) compared with the children in our study, in whom it was not possible to elicit a reflex at frequencies of less than 1Hz. A partial explanation of this is that the adults in the Burke and colleagues' study contained mainly spinal pathologies, including multiple sclerosis in six of the 18 cases. Tatto *et al.* (1985) reported that only 70 per cent of patients with increased tone, as a sequel to cortical or internal capsular vascular lesions, showed increased short-latency EMG activity. This finding and those of Burke and colleagues support the notion that non-electrical hypertonus is present in at least 30 per cent of adult strokes.

Conclusion

The implications for management are that, if the resistance to stretch is not due mainly to a velocity-dependent electrical reflex, drugs and neurosurgical management specifically aimed at reducing spasticity are clearly inappropriate. While the the treatment of hemiplegic children tends to be conservative, this cannot be said for the radical treatment options offered to diplegic children (Landau and Hunt 1990, Peacock and Staudt 1990). Systematic evaluation of reflex excitability will help to establish a more realistic perspective of its functional significance not only in the hemiplegias but also in other types of cerebral palsy.

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Appendix

Details of 14 hemiplegic children by case number, age, sex, side of hemiplegia, aetiology, associated childhood seizures, special educational needs, gait grade and muscle tone

Case	Age (yrs)[1]	Sex	Aetiology[2]		Side of hemiplegia	Seizures	Special education	Gait grade[3]	Tone			
			Congenital	Perinatal					Phasic spasticity	Tonic spasticity	Hemi–dystonia	Normal tone
4	11·7	M	IUGR		R			4	–	+	–	–
6	9·3	F	Silent		R			1	+	+	–	–
8	10·2	F		Asphyxia	R	+	+	2	–	–	–	+
10	6·4	M	Silent		R			1	–	+	–	–
11	6·7	M	Silent		R	+	+	4	–	+	–	–
12	11·3	F	Preterm		R			2	+	+	–	–
14	6·7	M	Silent		R			3	–	+	–	–
15	11·1	M	Silent		L			2	–	+	–	–
16	8·2	M	Preterm		R			4	+	+	–	–
17	6·2	M	Silent		R			4	+	+	–	–
18	12·0	M	IUGR		L			3	+	+	–	–
22	7·6	M		Asphyxia	L			3	+	+	–	–
23	12·2	F	Silent		L			1	–	–	+	–

[1]Mean (SD) = 9·4 (2·3).
[2]Silent = presumed silent infarct, IUGR = intra-uterine growth retardation; there was none with acquired hemiplegia.
[3]Gait score: 1 = heel strike, 2 = plantar strike, 3 = toe–heel strike, 4 = toe strike.

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Authors' Appointments
*J.-P. Lin, M.R.C.P., M.B., Ch.B., George Guthrie Research Fellow;
J. K. Brown, F.R.C.P.E., D.C.H., M.B., Ch.B., Consultant Paediatric Neurologist;
Department of Paediatric Neurology;
R. Brotherstone, E.P.T.A., Senior Neuropsychology Technician, Department of Clinical Neurophysiology;
Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF, UK.

**Correspondence to first author.*

SUMMARY
The quadriceps and hamstring muscles of 13 children with hemiplegic cerebral palsy were studied using ramp and sinusoidal stretches at three different muscle lengths. Overall, the hamstring muscles showed greater velocity sensitivity than the quadriceps, with the hamstrings having the lowest reflex velocity and frequency thresholds close to maximum knee extension. At this position, the hemiparetic hamstrings alone showed a weakly significant reduced reflex frequency threshold compared with non-paretic muscles. For all other muscle lengths, non-paretic and hemiparetic muscles displayed similar reflex thresholds when subjected to sudden, discontinuous or repetitive rhythmical, sinusoidal stretches. The number of muscles for which a reflex threshold could be established declined progressively as the angle at the knee joint approached 90° of flexion. Muscles for which no reflex threshold could be demonstrated had similar velocities and frequencies of stretch as those in which a reflex was obtained.

RÉSUMÉ
Evaluation de la spasticité dans la forme hémiplégique d'infirmité motrice cérébrale. I: Excitabilité réflexe proximale du membre inférieur
Les quadriceps et ischio-jambiers de 13 enfants hémiplégiques ont été étudiés à l'aide d'étirements en rampe et sinusoïde à trois longueurs musculaires différentes. Globalement, les ischio-jambiers présentaient une sensibilité à la vitesse plus grande que les quadriceps, la vitesse de réflexe et le seuil de fréquence les plus bas, près de l'extension maximale du genou. Dans cette position, les ischio-jambiers hémiparétiques présentaient seuls un seuil de fréquence de réflexe abaissé de façon faiblement significatif par comparaison avec les muscles non déficitaires. Pour toutes les autres longueurs de muscle, les muscles non déficitaires et hémiparétiques présentaient des seuils de réponse réflexe identiques lorsqu'ils étaient soumis à des étirements brusques, discontinus ou à répétition rythmique, ou sinusoïdaux. Le nombre des muscles pour lesquels un seuil de réflexe pouvait être établi baissait progressivement lorsque l'angle du genou approachait les 90° de flexion. Les muscles pour lesquels aucun seuil réflexe ne pouvait être mis en évidence, présentaient des vitesses et des fréquences d'étirement identiques à celles des muscles pour lesquels un réflexe était obtenu.

ZUSAMMENFASSUNG
Beurteilung der Spastik bei hemiplegischer Cerebralparese. I: Reflexverhalten im Bereich der proximalen unteren Extremität
Bei 13 Kindern mit hemiplegischer Cerebralparese wurden die Muskeln des Quadriceps, sowie die Mm. biceps femoris, semitendinosus und semimembranosus untersucht, indem bei drei verschiedenen Muskellängen ansteigende und sinusoidale Spannungen angewendet wurden. Insgesamt zeigten die Kniebeugemuskeln eine größere Geschwindigkeitssensitivität als der M. quadriceps, wobei sie die niedrigste Reflexgeschwindigkeit und Frequenzschwelle nahe der maximalen Kniestreckung hatten. In dieser Position zeigten nur die hemiparetischen Kniebeugemuskeln eine schwach signifikant verminderte Reflexfrequenzschwelle verglichen mit den nicht paretischen Muskeln. Bei allen anderen Muskellängen hatten die nicht paretischen und die paretischen Muskeln ähnliche Reflexschwellen wenn sie plötzlichen, unterbrochenen oder wiederholten rhythmischen, sinusoidalen Spannungen ausgesetzt wurden. Die Zahl der Muskeln, für die eine Reflexschwelle festgestellt werden konnte, nahm progressiv ab, je mehr sich der Kniegelenkswinkel einer Beugung von 90 näherte. Muskeln, bei denen keine Reflexschwelle nachgewiesen werden konnte, hatten ähnliche Spannungsgeschwindigkeiten und -frequenzen wie die, bei denen ein Reflex ausgelöst werden konnte.

RESUMEN
Evaluación de la espasticidad en la parálisis cerebral hemiplegica. Excitabilidad de los reflejos proximales de las extremidades inferiores
Se estudiaron los músculos cuádriceps y de la corva en 13 niños con parálisis cerebral hemiplégica utilizando estiramientos en declive y sinusoidales a tres longitudes diferentes del músculo. En conjunto los músculos de la corva mostraron una mayor velocidad de conducción que el cuádriceps con una velocidad refleja menor en los dinteles de frecuencia vecinos a la extensión maxima de la rodilla. En esta posición, los músculos paréticos de la corva mostraron un dintel de frecuencia refleja con una disminución ligeramente significativa, en comparación con los músculos no paréticos. Para todas las otras longitudes de músculo los no paréticos y los hemiparéticos mostraron unos dinteles reflejos semejantes cuando fueron sometidos a estiramientos súbitos, discontinuos o rítmicos, repetitivos y sinusoidales. El número de músculos para los que pudo establecerse un dintel reflejo disminuía progresivament a medida que el ángulo de la rodilla se acercaba a los 90° de flexión. Los músculos para los que no pudo obtenerse ningún dintel reflejo tenían unas velocidades i frecuencias de estiramiento símílares a las de aquellos en los que se obtuvo un reflejo.

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ASSESSMENT OF SPASTICITY IN HEMIPLEGIC CEREBRAL PALSY. II: DISTAL LOWER-LIMB REFLEX EXCITABILITY AND FUNCTION

Jean-Pierre Lin
J. Keith Brown
Ruth Brotherstone

It is commonly assumed that abnormal stretch reflexes are major contributors to the movement disorder in cerebral palsy. As previously discussed (Lin *et al.* 1993*a*), the elicited stretch reflex, along with voluntary and involuntary muscle activity, combines with the biomechanical properties of the muscle–tendon–joint complex to offer resistance to stretch. Many therapies (physical, pharmacological or surgical) are directed towards alleviating 'stretch reflex hypertonus' (see Fig. 8), though few studies have demonstrated a reduction in abnormal reflex activity or functional improvement after treatment. This lack of corroborative evidence is due in part to the difficulty of quantifying reflexes and reflex change. The problem is further compounded by the fact that not all EMG activity recorded is reflex, some being voluntary, some involuntary but mediated by descending supraspinal mechanisms, and some genuinely due to spinal and supraspinal stretch reflex mechanisms.

Many previous studies have recorded the clinical resistance felt on passive stretching according to the Ashworth scale (Ashworth 1964), which was originally devised to assess spasticity in patients with multiple sclerosis. Unfortunately it is not appropriate to use this non-linear and ordinal scale to diagnose spasticity because it merely grades resistance, irrespective of the factors producing it, and this may have led to a number of erroneous conclusions about treatment and outcome. Each component of the stretch resistance needs to be defined.

Although traditional tendon jerks are excellent diagnostic indicators of corticospinal damage, they are difficult to grade. They cannot measure spasticity quantitatively, first because they are produced by a stimulus which combines stretching and vibration (Lance and de Gail 1965), and second because the applied stimulus is not physiological, and provokes a supramaximal reflex response (all or nothing) as opposed to a graded one. What is needed is a clear functional definition of spasticity which can be used by any investigator. The clearest definition is that of Lance (1980): 'In both cerebral and spinal spasticity, the stretch reflex responses obtainable from extensor and flexor muscle groups of the upper and lower limbs increase approximately linearly with increase in the velocity of stretch. The reflex component of the increased tone may therefore be measured in terms of the threshold velocity required to evoke reflex activity and the slope of the EMG–velocity relationship.' This velocity dependence is seen repeatedly in the present study and forms the basis of

graded stretch reflex quantification.

A method of measuring the stretch reflexes of the muscles acting across the ankle joint is described for hemiplegic children, as a formal extension of clinical practice. The aim was to measure the reflex electromyogram (EMG) elicited, not the resistance felt (which would require other techniques). The velocity dependence of the reflex EMG in plantarflexor muscles is demonstrated. The reflex excitability on the non-paretic and hemiparetic sides is compared, and the potential clinical applications of assaying stretch reflexes are discussed in the wider context of all the cerebral palsies (see Figs. 8, 9).

Method

Population

A convenience sample of 14 children with congenital hemiplegia (mean age 9·3 (SD 2·2) years) were examined under controlled conditions. The clinical background of each child is summarized in Table I, which includes a brief description of the intervening therapy to which the child had been exposed. All the children had received 'eclectic' physiotherapy in the form of slow, passive stretching administered by the parents under six- to eight-weekly supervision of a professional physiotherapist from the time of diagnosis of the hemiparesis. Nine children had been using an ankle–foot orthosis (AFO) for between two and eight years, depending on age, and three of these also needed a night splint. One child had only had a night splint with physiotherapy. Five children proceeded to inhibitory casting: as one-off procedures over four to six weeks in three cases, twice in one case and yearly in another. One child subsequently underwent heel-cord tenotomy three years before the present assessment. In each of these cases, Table I shows the time interval between the onset of a given therapy and the time of reflex assessment.

Stretching the muscle

The stretch reflex can be specified if the position and speed of stretch of the muscle are known. How the muscle is stretched may also affect reflexes. The tibialis anterior (TA) and gastrocnemius–soleus (GS) muscle stretch reflexes were studied using two types of stretches: rhythmic and continuous sinusoidal stretches at increasing stretch frequencies, because they reduce the effects of acceleration and deceleration occurring between dorsiflexion and plantarflexion; and sudden ramp or discontinuous stretches at increasing angular stretch velocities, to test the muscles under conditions which mimic sudden displacements, such as landing on one's feet. These two types of stretch broadly represent the two main types of perturbation experienced at the ankle in everyday life. The children lay comfortably in the prone position with their feet over the end of a couch. Stretches were applied by the examiner without the aid of a torque motor; the advantage of this is that the stretches merely represented an extension of normal clinical practice with records of joint position and muscle activity. In order to stretch a muscle over a given range at a variety of speeds, the examiner varied the applied torque. Absolute limitations to speed of stretch were a combination of the inertial characteristics of the child's foot and the examiner's hand and arm (the inertia increasing with the square of the frequency of stretch), and the stiffness of the muscles resisting stretch. The torques applied would in many cases have exceeded several Newton metres (Nm). All tests were performed with the consent of the parents, who were present at all times.

Goniometry

The ankle–hindfoot displacement was measured in degrees with a Penny and Giles flexible goniometer (Figs. 1a to d). Displacement over time was monitored on a polygraph recorder. The examiner applied a displacement of 20° to 40° amplitude which included the position of resting plantarflexion (RPF, Fig. 1b) and the right-angle position (neutral, Fig. 1c) of the ankle joint.

EMG

Bipolar surface electrodes (Medicotest 'Blue Sensor') were placed parallel to the shaft of the leg with an inter-electrode distance of 6 to 10cm over the TA and

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TABLE I

Details of hemiplegic children by case number, age, sex, side of hemiplegia, congenital-perinatal or acquired aetiology, associated childhood seizures, special educational needs, gait grade, muscle tone and treatment. n = 14.

Case	*Age (yrs)*[1]	*Sex*	*Aetiology*[2]		*Side of hemiplegia*	*Seizures*	*Special education*	*Gait grade*[3]	*Tone*				*Treatment*[4]				
			Congenital	*Perinatal*					*Phasic spasticity*	*Tonic spasticity*	*Hemi-dystonia*	*Normal tone*	*Physio-therapy*	*AFO*	*Night splint*	*Teno-tomy*	*Inhib. cast.*
2	11·0	M	IUGR		R	+	+	4	+	+	–	–	10	6		3	6,4*
4	11·7	M	IUGR		R			4	–	+	–	–	7	2	3		Yearly
6	9·3	F	Silent		R			1	+	+	–	–	8				3
8	10·2	F		Asphyxia	R	+	+	2	–	–	–	+	9	8			
10	6·4	M	Silent		R			1	–	+	–	–	5				
11	6·7	M	Silent		R	+	+	4	–	+	–	–	8	1	4		
12	11·3	F	Preterm		R			2	+	+	–	–	10				
14	6·7	M	Silent		R			3	–	+	–	–	6	4			
15	11·1	M	Silent		L			2	–	+	–	–	2	2			
16	8·2	M	Preterm		R			4	+	+	–	–	6	2			
17	6·2	M	Silent		R			4	+	+	–	–	5	4·5			4
18	12·0	M	IUGR		L			3	+	+	–	–	11		2		
22	7·6	M		Asphyxia	L			3	+	+	–	–	7	5	5		1
23	12·2	F	Silent		L			1	–	–	+	–	11				

[1]Mean (SD) = 9·3 (2·2).
[2]Silent = presumed silent infarct; IUGR = intra-uterine growth retardation; there was none with acquired hemiplegia.
[3]Gait score: 1 = heel strike, 2 = plantar strike, 3 = toe–heel strike, 4 = toe strike.
[4]Years intervening between treatment and assessment; AFO = ankle–foot orthosis; inhib. cast. = inhibitory casting.
*Six and four years before assessment.

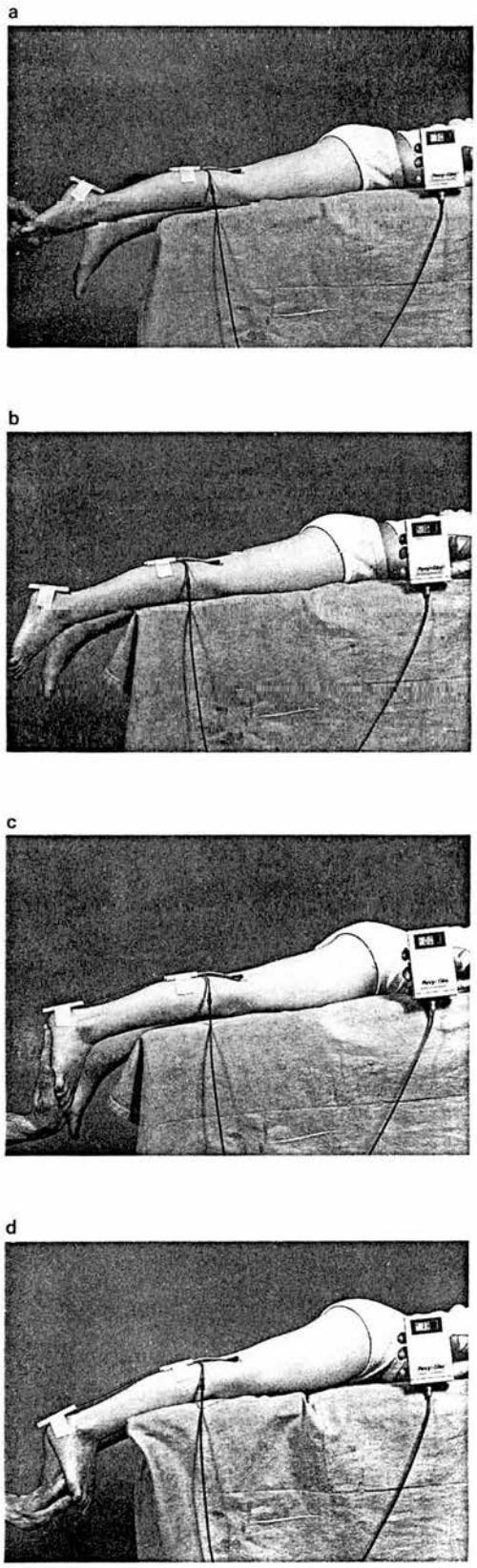

Fig. 1. Six-year-old boy with congenital right hemiparesis (case 17) lies prone on couch with Penny and Giles twin-axis goniometer measuring hindfoot angular displacements. (a) Maximum passive plantarflexion, (b) resting plantarflexion, (c) 'neutral' position (90°), and (d) maximum passive dorsiflexion. Passive joint range = d minus a; effective range = d minus b (see Table II).

GS muscles, depending on the size of the child. The EMG bandwidth was set at 0·5 to 150Hz and stretches were imposed on initially resting muscles. All polygraphic recordings show a time signal, dorsiflexor (TA) EMG in μV, ankle displacement (GON) in degrees and the plantarflexor (GS) EMG in μV. By convention, dorsiflexion (DF) is upwards, indicated by an arrow, and plantarflexion (PF) is downwards for sinusoidal and ramp stretches (see Fig. 2). In addition to the stretch reflexes, we measured resting plantarflexion (RPF) and the passive joint ranges, together with the maximum voluntary isometric EMG of the plantarflexor and dorsiflexor muscle groups. The maximum speed of voluntary alternating plantarflexion/dorsiflexion (PF/DF) at the ankle was recorded in Hertz (Hz) as a measure of ankle dexterity, together with the amplitude of voluntary movement in degrees. Ankle and IA jerks were also recorded, along with the number of beats of ankle clonus elicited.

Statistics and analysis

The results for hemiparetic and non-paretic limbs were expressed in terms of the reflex frequency threshold and reflex frequency gain for sinusoidal stretches, and reflex velocity threshold and reflex velocity gain for ramp stretches. Reflex EMG thresholds were derived from the intercept of a linear regression equation of the EMG in μV plotted against either frequency or velocity of stretch. The slope of the regression equations defined the reflex EMG gain. Since the data for each limb were gathered over time, variations in mental state due to the influence of non-specific stimuli were averaged over the same period and the regression equations for each limb and each mode of stretch were thus specified in terms of reflex thresholds and gains which

TABLE II

Passive and active hindfoot ankle goniometry and ankle dexterity of non-paretic and hemiparetic limbs

Ankle position[1]	*Non-paretic limb*[2]		*Hemiparetic limb*[2]		*Paired t test (p value)*
	Lower	*Upper*	*Lower*	*Upper*	
Passive PF (N = 14)	48·5°	58·3°	48·2°	59·5°	NS
Resting PF (N = 12)	68·5°	80·1°	68·9°	75·3°	NS
Passive DF (N = 14)	101·6°	108·9°	93·0°	100·4°	<0·0002
Passive range (N = 14)	46·5°	57·1°	35·5°	50·0°	<0·02
Effective range (N = 12)	24·2°	36·5°	18·5°	30·3°	NS
Active range (N = 13)	19·2°	35·2°	2·7°	13·6°	<0·0003
Ankle dexterity (N = 13)	1·5Hz	2·8Hz	0·2Hz	1·0Hz	<0·0002

[1]PF = plantarflexion; DF = dorsiflexion; passive range = passive dorsiflexion minus passive plantarflexion; effective range = passive dorsiflexion minus resting plantarflexion; ankle dexterity = frequency of voluntary alternating plantarflexion and dorsiflexion.
[2]99 per cent confidence intervals.
90° = foot at right angle to tibia. A joint angle of >90° indicates dorsiflexion and a joint angle of <90° indicates plantarflexion beyond neutral.

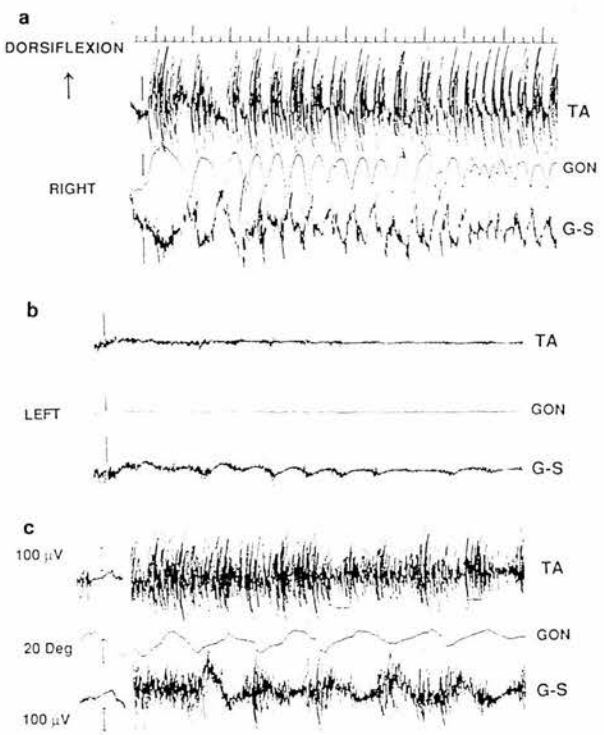

Fig. 2. (a) *Repetitive voluntary alternating dorsiflexion/plantarflexion of non-paretic ankle of a 12-year-old boy with congenital left hemiparesis (case 18): note that modulation of frequency and amplitude of movement* (GON) *is accompanied by a typical triphasic EMG response of tibialis anterior* (TA) *and gastrocnemius-soleus* (G-S) *muscles. Dorsiflexion is upward in all traces.* (b) *By contrast, hemiparetic limb shows little alternating movement at ankle and is accompanied by amorphous co-contraction of* TA *and* G-S *muscles.* (c) *Hemidystonic limb of 12-year-old girl with congenital hemidystonia (case 23) shows large-amplitude continuous co-contraction:* G-S *is more modulated than* TA *muscle which appears more dystonic.* Large divisions = *1s.*

represent the 'reflex excitability' of the limbs at that time. Data are tabulated in the form of 99 per cent confidence intervals. Hemiparetic and non-paretic reflex thresholds, reflex gains and ankle dexterity measures were compared for each case using paired *t* tests, as were the passive ankle-joint ranges obtained.

Results

Passive ankle goniometry

Table II shows the maximum impositions of passive plantarflexion, resting plantarflexion, passive dorsiflexion and passive joint-ranges of hemiparetic and non-paretic limbs, and Figure 1 shows typical joint positions. The angles of passive and

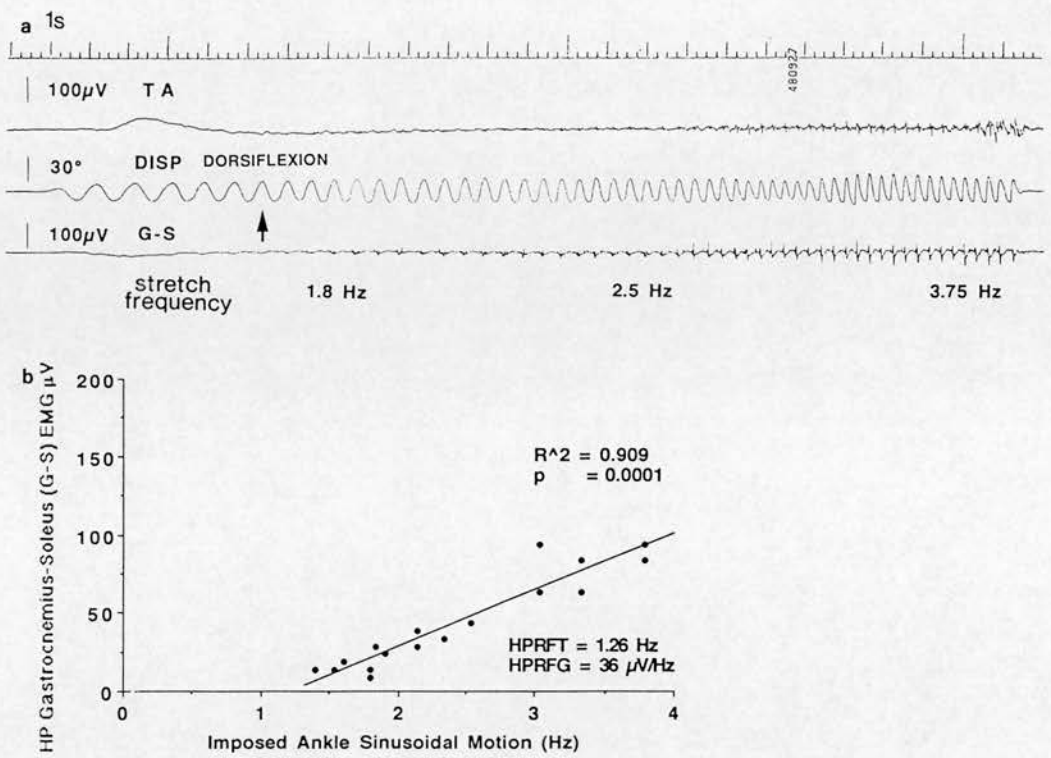

Fig. 3. (a) Sinusoidal stretch reflex excitability. Sinusoidal stretching of hemiparetic muscles of eight-year-old boy (case 16) with congenital right hemiparesis secondary to preterm birth (but no intraventricular haemorrhage). Reflex EMG threshold response (reflex frequency threshold) is seen at 1·8Hz for G-S *muscles and this increases with increasing frequency of stretch.* TA *reflex frequency threshold is just over 2Hz and* TA *EMG also rises with increasing stretch frequency (maximum 3·75Hz).* DISP = *ankle displacement. (b) Sinusoidal stretch reflex excitability curve. Reflex EMG for hemiparetic* G-S *muscles in (a) is plotted against frequency of sinusoidal stretch. There is a highly linear regression between reflex EMG and frequency of stretch. Intercept of linear regression equation gives hemiparetic reflex frequency threshold (*HPRFT*), and slope of regression line is measure of reflex frequency gain (*HPRFG*). (See Fig. 2 for explanation of other abbreviations.)*

resting plantarflexion between non-paretic and hemiparetic limbs did not differ statistically. All ankle joints could be passively dorsiflexed to beyond 90° (neutral), non-paretic limbs more so than hemiparetic ones ($p<0·0002$). The overall passive joint ranges were greater on the non-paretic side by about 8° ($p<0·02$), but the effective joint ranges between the resting position and maximum passive dorsiflexion showed no statistical difference.

Voluntary ankle dexterity and amplitude of movement

A typical comparison of the difference in ankle dexterity between affected and unaffected limbs can be seen in Figure 2, which shows the voluntary ankle movements and corresponding voluntary dorsiflexor (TA) and plantarflexor (PF) muscle EMG activity. For a 12-year-old boy with congenital left hemiplegia, the non-paretic limb (Fig. 2a) was able to vary the frequency and amplitude of movement, and the surface EMG recordings show a typical 'triphasic' pattern of activity in which agonist and antagonist alternate to allow motion. On the hemiparetic side (Fig. 2b), a low-grade co-contraction of opposing muscles results in almost complete loss of movement. Figure 2c shows the dexterity for one subject with congenital hemidystonia, in which the attempted alternating movements of the hemidystonic limb resulted in large co-contractions of TA and GS muscles with loss of the normal triphasic response. This dystonic pattern fluctuates rapidly,

TABLE III
Reflex velocity threshold and allow for non-paretic and hemiparetic gastrocnemius–soleus (G-S) muscles in response to ramp stretches

	N	*Non-paretic limb*[1]		*Hemiparetic limb*[1]		*Paired t test*
		Lower	*Upper*	*Lower*	*Upper*	
Reflex velocity threshold (°/s)	12	−144·3	90	−192·3	114·3	NS
Reflex velocity allow (μV/°/s)	12	0·257	0·581	0·303	0·908	NS

[1]99 per cent confidence intervals.

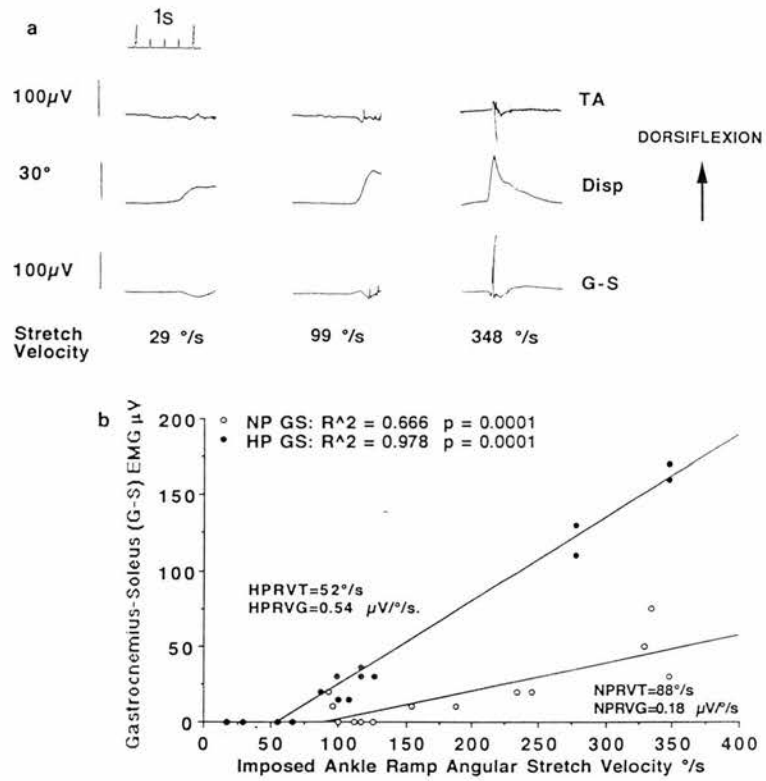

Fig. 4. (a) *Ramp stretch reflex excitability. Hemiparetic ankle of 10-year-old boy with perinatal hemiparesis (case 8) is rapidly dorsiflexed at increasing angular velocities. Stretch reflex of* G-S *muscles is seen at 99°/s and has increased considerably at 348°/s, but is absent at 29°/s.* (b) *Ramp stretch reflex excitability curve. Reflex EMG for hemiparetic limb shown in* (a) *is plotted against ramp stretch angular velocity at ankle, along with data from non-paretic limb. Both limbs show highly significant regression of reflex EMG with velocity of stretch.* NP = *non-paretic,* HP = *hemiparetic,* RVT = *reflex velocity threshold,* RVG = *reflex velocity gain.*

allowing brief periods of relatively good dexterity interspersed with bursts of co-contraction which abolish movement. Ankle dexterity and amplitude of voluntary motion were greatly reduced for hemiparetic limbs ($p<0·0002$ and $p<0·0003$, respectively; Table II). In five cases the hemiparetic limb had no dexterity at all (*i.e.* a hemiparetic ankle dexterity of 0Hz), and in the remaining eight the mean hemiparetic ankle dexterity was 1·1Hz. The relationship between the measures of dexterity and reflex excitability are examined below.

Reflex excitability

Regardless of whatever sinusoidal or ramp stretches were used, non-paretic and hemiparetic leg muscles were initially silent until a critical threshold speed of stretch was achieved. This was termed the reflex threshold. With increasing speeds the non-paretic reflex velocity gain of sinusoidal or ramp stretch, muscles

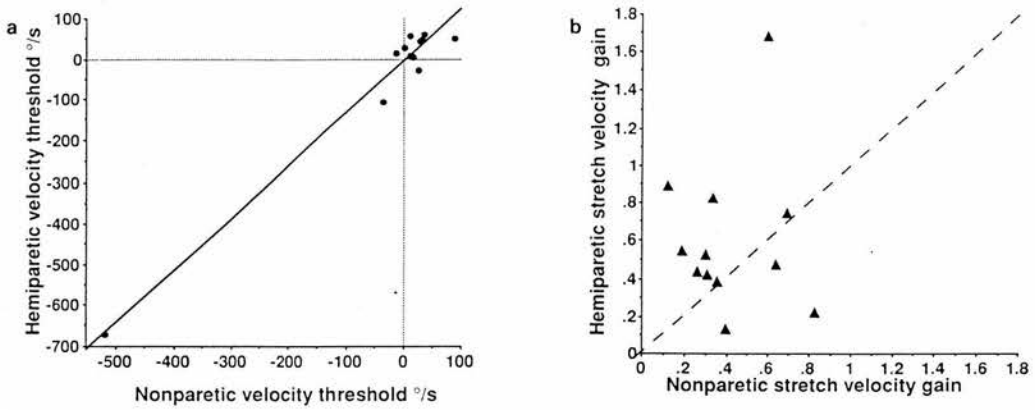

Fig. 5. (a) Reflex velocity thresholds (RVT) for 12 NP and corresponding HP limbs are plotted against each other and fall roughly equally about identity regression line. Note that negative RVT indicated muscle pre-activation in 4/12 cases. Overall, NP *and* HP *limbs had a similar RVT on paired t testing (Table III)). (b) Reflex velocity gains (RVG) of 12 NP and corresponding HP limbs plotted against each other, showing wide scatter about identity line (*broken*): there was no statistical increase of RVG for HP compared with NP limbs (Table III).*

showed a strong linear increase in reflex EMG, and this velocity-dependent increase was referred to as the reflex gain. Reflex frequency, velocity thresholds and gains were obtainable from the intercepts and slopes of the individual regression equations for most limbs (Table III). Such threshold and gain parameters reflect the reflex excitability of the motor system.

Sinusoidal stretch

The reflex EMG against frequency of sinusoidal stretch for the hemiparetic limb of case 16, an eight-year-old boy with a congenital right hemiplegia, is shown in Figure 3a and a plot of EMG amplitude against frequency in Figure 3b. The first EMG reflex activity occurred at just over 1Hz and increased linearly with faster frequencies of stretch. The intercept of the linear regression equation gave a derived reflex frequency threshold of 1·2Hz. The slope of the regression line is a measure of the reflex frequency gain in μV/Hz.

Ramp stretch

The effect of imposing a sudden, discontinuous ramp stretch on the calf muscles of the hemiparetic limb of case 16 is shown in Figure 4a, while the results for hemiparetic and non-paretic limbs are plotted graphically as EMG against stretch velocity in Figure 4b. Note that each limb had a reflex velocity threshold derived from the intercept of the linear regression equation: this was 52°/s for the hemiparetic and 88°/s for the non-paretic limb. There was a also a linear increase in EMG with increasing stretch velocity, and the reflex velocity gain (as given by the slope of the regression equation) was 0·54μV/°/s for the hemiparetic and 0·18μV/°/s for the non-paretic limb. In this case the hemiparetic limb was more excitable than the non-paretic limb, exhibiting a lower-velocity threshold and greater gain.

Comparative analysis

The responses to ramp stretches of the gastrocnemius–soleus muscles were analysed in detail, and Table III shows the 99 per cent confidence intervals for the reflex velocity threshold and reflex velocity gain of the non-paretic and hemiparetic limbs of 12 children. Despite apparent differences between non-paretic and hemiparetic limbs in individual cases, paired *t* testing of reflex velocity thresholds and gains were not statistically different for the group as a whole. Four subjects had a negative reflex velocity threshold in one or both limbs, which corresponds to muscle pre-activation during stretch (Fig. 5a). The paired reflex velocity gain for these 12 subjects is

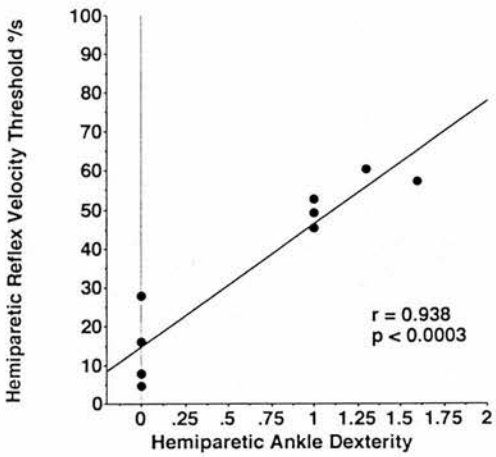

Fig. 6. Ankle dexterity and RVT. When RVT of nine hemiparetic HP limbs is plotted against ankle dexterity in Hz, two groups of HP limbs are identified: more or less plegic with low RVT (<40°/s) and paretic (RVT >40°/s), indicating that dexterity diminishes as RVT increases.

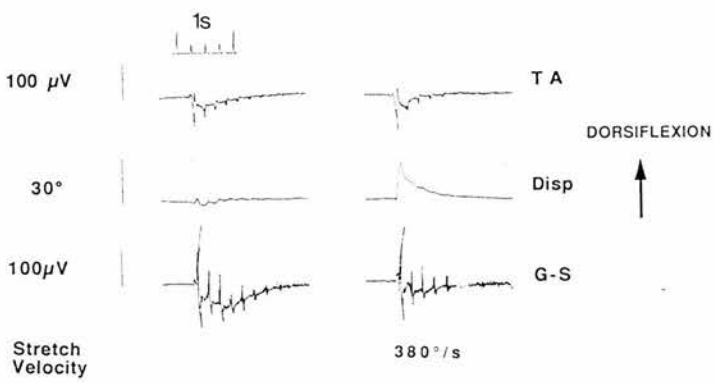

Fig. 7. Clonus appears to be independent of velocity of ramp stretch in 12-year-old boy with left hemiparesis secondary to intra-uterine growth retardation (case 18), and was a variable finding. (See previous figures for explanation of abbreviations.)

plotted in Figure 5b, showing that the hemiparetic reflex velocity gain was greater than the non-paretic reflex velocity gain in seven cases, equal to it in two cases and lower than it in three cases.

Reflex excitability and dexterity

Ankle dexterity frequencies for non-paretic and hemiparetic sides are given in Table II as the maximum frequency of voluntary alternating plantarflexion/dorsiflexion. The hemiparetic limb had a significantly lower dexterity frequency than the companion non-paretic limb (non-paretic ankle dexterity 2Hz, hemiparetic ankle dexterity 0·6Hz, $p<0·0002$). The hemiparetic reflex velocity threshold and ankle dexterity were highly correlated for nine cases ($r=0·938$, $p<0·0003$), with hemiparetic limbs falling into two groups: (1) totally plegic (mean ankle dexterity 0Hz) with low reflex velocity threshold of <40°/s, and (2) partially functional hemiparetic limbs (mean ankle dexterity = 1·15Hz) with a reflex velocity threshold of >40°/s (Fig. 6). This suggests that there is an inverse relationship between function and reflex excitability: the lower (*i.e.* the more excitable) the reflex velocity threshold,

the lower the voluntary function. There was no association between reflex velocity gain and ankle dexterity.

Clonus

Clonus appeared to be independent of the velocity of stretch (Fig. 7). The presence of clonus was not a consistent finding for hemiparetic limbs and so could not be used to grade reflex excitability.

Discussion

Controlled sinusoidal or ramp stretches at physiological rates akin to those normally encountered at the ankle for walking (ankle angular velocity of 20°/s; Hufschmidt and Mauritz 1985), running and jumping (ankle angular velocities of 295 to 410°/s; Dietz 1981), can provide a graded reflex excitability curve from which the behaviour of the reflexes can be specified in terms of a reflex threshold and reflex gain. Ankle goniometry showed a 5° to 10° restriction in maximum joint-range on the hemiparetic side, and all hemiparetic limbs were clearly abnormal, as established by the ankle dexterity test ($p<0{\cdot}0002$). Clear differences in reflex excitability between non-paretic and hemiparetic limbs were not demonstrated, though voluntary ankle dexterity on the hemiparetic side appears to be inversely related to reflex excitability.

These data suggest that, for the most part, congenital hemiplegia is not accompanied by marked differences in reflex excitability between non-paretic and hemiparetic sides. Although the children included represent a heterogeneous group of hemiplegic children (Lin and Brown 1992), it is questionable whether the term 'spastic hemiplegia'—with its implication of a high degree of reflex excitability—is justified in such a group, especially since it is widely held that the commonest forms of cerebral palsy are spastic in nature. As indicated in Table I, a variety of stretching treatments applied more or less continuously with varying enthusiasm by the parents, and the individual child's degree of compliance, may have influenced the natural history of the stretch reflex excitability in this group, an issue which can be resolved only by prospective evaluation (Figs. 8 and 9). The question

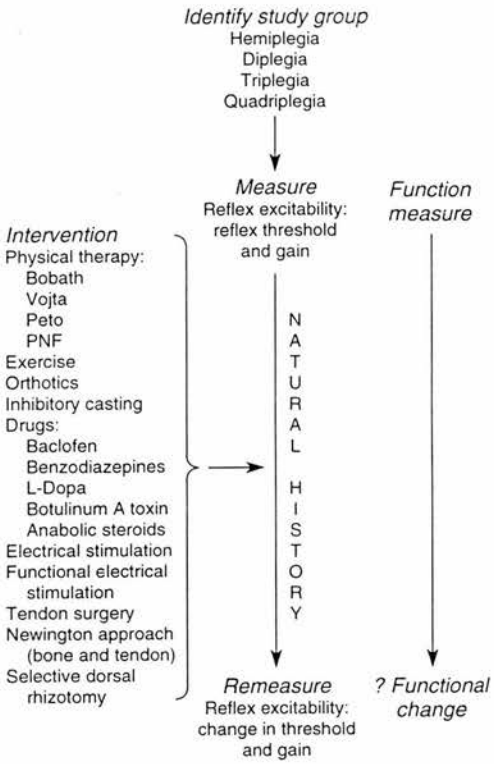

Fig. 8. Model for measurement of spasticity. After identifying study group according to distribution of neurological signs, reflex excitability (RVT and RVG) and function (e.g. dexterity) are measured before intervention. Several possible therapies are listed on left. After treatment, reflex excitability and function are remeasured. Treatment may have no effect on reflex excitability or function, or may improve function without affecting stretch reflexes, or both reflex excitability and function may improve with treatment. Model offers means of assessing patient and effectiveness of treatment.

of spasticity in children with cerebral palsy is even more pressing, given the growing popularity of radical procedures such as selective dorsal rhizotomy in children with diplegia.

What alternative explanations are there for the resistance felt on stretching calf muscles or other muscles of the paretic limb? Herman (1970) demonstrated the natural history of reflex excitability in adults following strokes. In his large study, four groups were identified according to the length of time since the cerebral insult. Excitability was depressed in the aftermath of the injury in group I, gradually increasing to a peak in groups II and III, and frankly depressed in group IV, which he called the 'pre-contracture

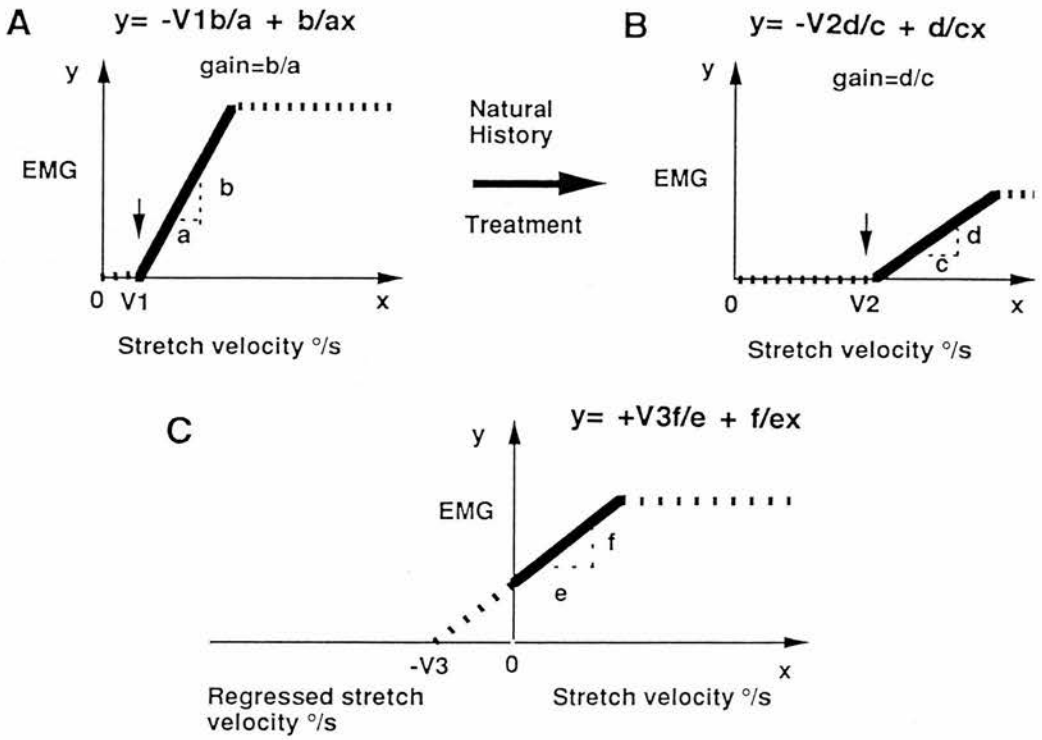

Fig. 9. Effect of natural history, treatment or muscle pre-activation on reflex excitability, where V1 *and* V2 *and* −V3 *are hypothetical reflex velocity thresholds, and* −V3<V1<V2. *For* A *and* B, *stretch reflex velocity gain is indicated by slopes of reflex excitability curves,* b/a *and* d/c, *where* d/c<b/a. *Graded increase in reflex EMG with velocity of stretch is indicated by* solid lines *and corresponding linear equation is written above each curve.* Broken lines *indicate that response to stretch is sigmoid with little or no EMG until a critical velocity threshold (*V1, V2*) followed by a rise in reflex EMG with stretch velocity and, finally, a plateau (motor unit pool maximally recruited). (N.B. If muscle is active before stretch (at 0 stretch velocity), stretch threshold is negative (*−V3*). Such a pre-activation may increase gain of stretch reflex.)*

group'. All our subjects had suffered either prenatal or perinatal brain injuries and had lived with their brain injuries for between six and 12 years. Growth, development and the potential for recovery in the form of cerebral plasticity may have reduced the reflex excitability in our group, emphasising the difference between childhood and adult strokes. Recent studies have indicated that children with cerebral palsy (CP) may retain a degree of reciprocal excitation which is usually lost or suppressed by the developing intact brain (Gottlieb *et al.* 1982, Myklebust 1990). Opinions vary on the extent to which reciprocal inhibition is lost or preserved in CP. Loss of reciprocal inhibition following H reflex testing has been cited as a possible explanation for involuntary co-contraction during voluntary tasks at the ankle in children with CP (Leonard *et al.* 1990). Increased reciprocal inhibition has been found to determine the probability of single motor units being fired from the tibialis anterior muscle after stimulation of the posterior tibial nerve of 15 adults who had had CP since birth (Berbrayer and Ashby 1990). The presence of co-contractions during voluntary dexterity tasks was seen in all our subjects with hemiplegia on the hemiparetic side, reflecting a loss of direct cortical control over the spinal motor neuron pool.

A number of studies have said that changes in the plastic properties of muscle may account for an increase in non-electrical resistance to stretch in calf muscles of adults following strokes (Herman 1970, Hufschmidt and Mauritz 1985) and of children with CP (Foley 1961, Dietz *et al.* 1981, Dietz and Berger 1983). We have suggested that a clear distinction must be made between the

characteristic hemiposture which often comes and goes with intention, and the phenomenon of spasticity which depends on muscle stretch (Lin and Brown 1992). The hemiplegic syndrome of childhood comprises at least four distinct entities: abnormal motor planning, abnormal limb postures, a variable degree of reflex excitability and plastic muscle changes. Such plastic properties of muscle are very prominent in hemiparetic limbs in the form of muscle creep, and are the subject of another study which is currently in progress. Posture, reflex excitability and plastic muscle change will all offer resistance to muscle stretch and contribute to what is refered to clinically as 'mucle tone'.

Neuro-anatomical basis for spasticity

Previous attempts at mapping out the neuro-anatomical correlates of spasticity have yielded surprising results in primate models. Tower (1940) first demonstrated that bilateral division of the pyramids in the cat or monkey did not produce spasticity, a finding widely supported by other experiments and by clinical observations in man (Tasker 1980). Further studies implicated lesions of the supplementary motor area 4s in the pathogenesis of spasticity, but more recent studies in squirrel monkeys failed to demonstrate onset of spasticity six months following lesions to unilateral primary motor cortex (area 4) or unilateral or bilateral supplementary motor cortex (area 4s) alone. Only after bilateral lesions to the primary motor cortex (area 4) was it possible to demonstrate spasticity (Tasker 1980), which reinforces the clinical observation that spasticity is a strong correlate of bilateral damage to the motor cortex. This evidence, together with the known influence of time in reducing reflex excitability (Herman 1970), may further explain why children with congenital hemiparesis, tested some six to 12 years after sustaining cerebral damage, exhibit relatively little stretch reflex excitability.

Conclusion

The fundamental clinical importance for distinguishing between muscle plasticity and spasticity can be understood in relation to the therapies available for managing children with CP illustrated in Figure 8. It would be inappropriate to concentrate treatment on reducing muscle excitability if in fact there is little reflex excitability to be elicited. A recent attempt at measuring stretch reflex excitability in the proximal lower-limb muscles (Lin *et al.* 1993) supports the current findings that lower-limb stretch reflexes are not particularly exaggerated in congenital hemiplegia. The scheme in Figure 8 offers a framework for assessing children and available treatments or simply following up the natural developmental history without intervention.

If spasticity (in the form of a velocity-dependent increase in stretch reflex excitability) requires treatment, the therapist should demonstraate a change in reflex excitability, *i.e.* a higher reflex velocity threshold and lower reflex velocity gain (Fig. 9). If spasticity is thought to interfere with function, funcional improvement following a reduction in reflex excitability must be shown. The individual assesment of children in this way presents an enormous task for all those involved in the management of CP, and it is likely to be some time before such systematic assessments are widely available. Measurements and imaging form the basis of rational assessment and treatment in every other field of medicine and are no less applicable to the difficult field of motor disorders, which requires attention to the different components contributing to abnormal muscle tone, posture and movement.

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Authors' Appointments

*J.-P. Lin, M.R.C.P., M.B., Ch.B., Edinburgh University, Faculty of Medicine, George Guthrie Research Fellow;
J. K. Brown, F.R.C.P.E., D.C.H., M.B., Ch.B., Consultant Paediatric Neurologist;
Department of Paediatric Neurology;
R. Brotherstone, E.P.T.A., Senior Neurophysiology Technician, Department of Paediatric Neurophysiology;
Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF, UK.

**Correspondence to first author.*

SUMMARY
A clinical method for measuring the stretch reflex theshold and gain of muscles acting across the ankle joint in children with congenital hemiplegia is described. The stretch reflexes of all limbs were velocity-dependent. Hemiparetic limbs were not necessarily spastic compared with non-paretic limbs, suggesting that the term 'spastic hemiplegia' should be used more selectively and emphasis be placed on the heterogeneity of the hemisyndromes of childhood. Abnormal motor control, planning and dexterity, the hemipostures and plastic (non-electrical) muscle changes may be more important measures of impairment than reflex excitability. A general scheme for assessing reflex excitability and available treatments applicable to all types of cerebral palsy is proposed.

RÉSUMÉ
Evaluation de la spasticité dans l'IMC hémiplégique. II: Excitabilité réflexe et fonction de la partie distale des membres inférieurs
Une méthode clinique pour mesurer le seuil du réflexe d'étirement et la force des muscles agissant sur la cheville chez les enfants présentant une hémiplégie congénitale est décrite. Les réflexes d'étirement de tous les muscles des membres dépend de la vitesse. Les membres hémiparétiques n'apparaissent pas nécessairement spastiques par comparaison avec les muscles non parétiques, ce qui conduit à suggérer que le terme 'hémiplégie spastique' devrait être utilisé plus sélectivement et il faudrait insister sur l'hétérogénéité des hémisyndromes de l'enfance. Un contrôle moteur anormal, la qualité de la prévision et de la dextérité, les hémipostures et les modifications plastiques des muscles (non électriques) peuvent être des mesures plus importantes de l'incapacité que l'excitabilité réflexe. Un schème général pour apprécier l'excitabilité réflexe et les traitements utilisables, applicables à tous les types d'IMC, est proposée.

ZUSAMMENFASSUNG
Beurteilung der Spastik bei hemiplegischer Cerebralparese. II: Reflexexcitabilität und -funktion im distalen Bereich der unteren Extremität
Es wird eine klinische Methode zur Messung der Streckreflexschwelle und der Muskelkraft im Bereich des Fußgelenkes bei Kindern mit kongenitaler Hemiplegie beschrieben. Alle Streckreflexe sind geschwindigkeitsabhängig. Hemiparetische Extremitäten sind nicht notwendigerweise spastisch, verglichen mit nicht-paretischen Extremitäten, d.h. der Ausdruck 'spastische Hemiplegie' sollte selektiver benutzt und die Verschiedenartigkeit der Hemisyndrome im Kindesalter sollte stärker hervorgehoben werden. Gestörte motorische Kontrolle, Ausführung und Geschicklichkeit, Hemisymptomatologie und elastische (nicht elektrische) Muskelveränderungen sind wahrscheinlich wichtigere Parameter für eine Schädigung als die Reflexerregbarkeit. Es wird ein allgemeines Schema zur Beurteilung der Reflexerregbarkeit und der verfügbaren Behandlungsmethoden vorgestellt, das auf alle Arten der Cerebralparese anwendbar ist.

RESUMEN
Evaluación de la espasticidad en el parálisis cerebral hemiplégica. II: Excitabilidad y función refleja distal en la extremidad inferior
Se describe un método clínico para medir el dintel del reflejo de estiramiento y su ganancia en los músculos que actúan a nivel de la articulación del tobillo en niños con hemiplegia congénita. El reflejo de estiramiento en todas las extremidades depende de la velocidad. Las extremidades hemiparéticas no son necesariamente espásticas en comparación con los miembros no paréticos, lo que sugiere que el término 'hemiparesia espástica' debería usarse más selectivamente y dar importancia a la heterogeneidad de los hemisíndromes en la infancia. El control motor anómalo, la planificación y la destreza, las hemiposturas y los cambios musculares plásticos (no eléctricos) pueden ser las mediciones más importantes de la alteración, más que la excitabilidad refleja. Se propone un esquema general para evaluar la excitabilidad refleja y los posibles tratamientos aplicables a todos los tipos de parálisis cerebral.

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diagnosis of diabetes, is that they did not develop insulin dependency acutely. We will follow-up these women to see how many will ultimately be classified as having IDDM.

Many women with GDM will later develop frank diabetes, and it has been estimated that about 2% of women with GDM will proceed to IDDM during the subsequent 15 years.[28] In the present study, 5% of all women with GDM were anti-GAD positive, and most of the anti-GAD-positive women with GDM (5 of 6) required insulin treatment during pregnancy. How many of these anti-GAD-positive women will later develop IDDM remains to be seen. Identification of women with GDM who have a high likelihood of developing IDDM is important to allow early intervention to prevent IDDM or its complications. Whether the predictive value of anti-GAD for IDDM is different in women and men is not known. In a study of recent-onset IDDM in childhood in Australia, there was a small sex difference in percentage of anti-GAD positives (females 75%, males 63%; C F Verge, Royal Alexandra Hospital for Children Sydney, Australia, personal communication). There is no evidence to suggest that pregnancy should influence anti-GAD concentrations—in our study anti-GAD-positive women who were retested serially over several pregnancies had stable values of anti-GAD.

The sensitivity and specificity of anti-GAD positivity to detect IDDM in sera from pregnant women were high. Results were sufficiently good for the anti-GAD assay to be used prospectively as a screening test to detect individuals at risk of developing IDDM. On the other hand, before mass screening can be recommended, an effective and acceptable treatment to prevent IDDM should be available. Furthermore, it is important to find out how much beta-cell capacity is left in subjects who are anti-GAD positive. Experimental data indicate that loss of about 80–90% of beta cells is needed before clinical symptoms or diabetes occur.[29] If anti-GAD positivity is present when people are approaching the critical level of beta-cell capacity, anti-GAD positivity may be used for secondary and tertiary prevention of IDDM.

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Physiological maturation of muscles in childhood

J-P Lin, J K Brown, E G Walsh

Summary

Little is known about the motor muscle physiology of growing children and it has been assumed that increasing motor dexterity and speed correspond solely to brain maturation, although there is evidence that muscles mature from studies of other mammalian species. Such a finding in children would throw light on the possible mechanics of motor delays in speech, hand function, athletic, and musical skills as well as helping towards a better understanding of pathological motor disorders such as the cerebral palsies.

Our non-invasive observations of soleus-muscle relaxation times in healthy children suggest that a child's muscles are initially slow to relax but relaxation doubles in speed up to adult rates by early adolescence. Half-relaxation times halved from about 90 ms at age 3 to about 40 ms at age 10. On the basis of these studies, we suggest that muscle maturation rate-limits motor tasks; which should be borne in mind when considering motor development in muscles and when attempting to determine the effects of early brain or spinal cord damage.

Such methods of measuring the dynamics of muscle open up new areas of research and offer the possibility of developing operationally-defined in-vivo measurements for use in motor assessment and for following up the effects on muscle or treatments such as physiotherapy, orthotics, surgery, and drugs.

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Paediatric Neurology, Royal Hospital for Sick Children, Sciennes Road, Edinburgh (J-P Lin MRCP, J K Brown FRCPE) **and Department of Physiology, University of Edinburgh, Edinburgh, UK** (E G Walsh FRCPE)

Correspondence to: Dr J-P Lin, Paediatric Neurology, Great Ormond Street Hospital for Children, London WC1N 3JH, UK

Introduction

This study was undertaken as a background investigation into the contractile properties of muscles in children with cerebral palsy. Current treatments for improving posture and mobility in children with cerebral palsy, with the exception of selective dorsal rhizotomy, focus on loading muscles with physiotherapy, orthotics, exercise, or electrical stimulation; or unloading muscles with soft tissue releases, immobilisation, or botulinum toxin. Despite widespread use of such treatments, little is known about the physiology or pathophysiology of the muscles so treated; indeed, little is known about the natural history of muscle in the context of the cerebral palsies.

Human motor development studies have concentrated largely on observations of the maturation of the nervous system,[1,2,3] whilst muscles are considered to be developmentally static, which is surprising considering in-vitro information on the development and adaptability of muscle drived from other mammals[4,5] which began when Buller and Eccles showed that a committed muscle-fibre type could be transformed from slow-twitch to fast twitch and vice versa in their cross-innervation experiments,[6] confirming that impulse traffic down the nerve conditions the fibre-type.[6]

Human muscle-fibre diameter increases with age; clear sex differences in diameter emerge around the age of 10 years,[7] and muscles grow and become stronger with age.[8,9] That so little else is known about maturation of human skeletal muscle stems partly from the fact that muscle physiology has relied on methods requiring biopsy specimens or the use of electrical stimulation to obtain motor-unit contractions followed by muscle biopsy for myosin-ATPase studies of fibre type.[10] Neither muscle biopsy nor repetitive electrical stimulation of healthy muscles are suitable approaches in children.

We have developed a painless, non-invasive technique for measuring the effects of age on the relaxation of calf muscle in healthy children.

Methods

With approval from the Lothian Health Board Paediatric and Reproductive Medicine Research Sub-Committee and parental consent, we investigated the influence of muscle length and age on the contractile properties of soleus muscles in 22 healthy children aged 3 to 10 years with an instrument previously described for use in adults[11,12] but modified to take into account variation in foot size with age (figure 1). Results from children were compared with those from 176 healthy young adults, 91 females and 85 males, mean age 18·5 years.

With the subject relaxed and lying on the left with the knee flexed to 90° to eliminate the influence of the gastrocnemius muscle, soleus muscle twitches were generated by a single Achilles tendon tap which caused a monosynaptic reflex muscle-twitch contraction. Disposable surface electrodes 10 cm apart overlying the soleus were used to record electromyographic (EMG)

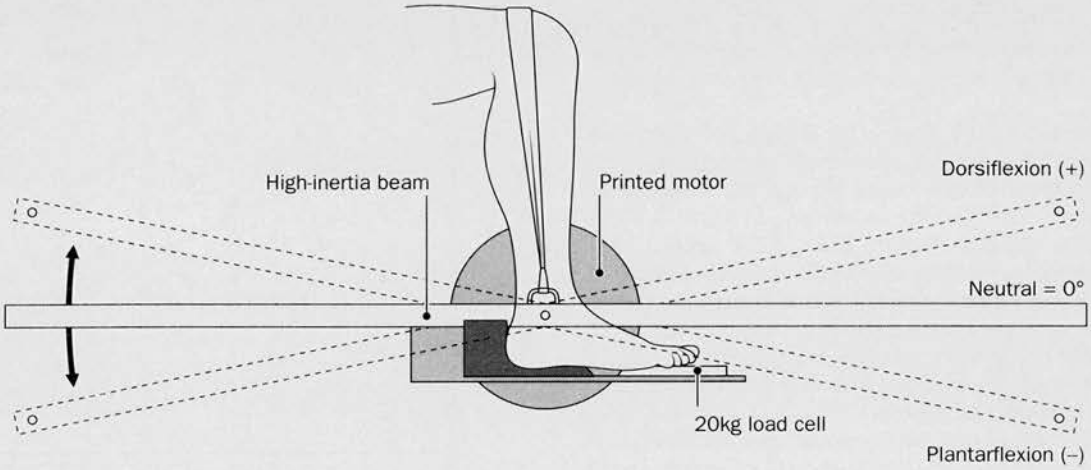

Figure 1: **Measurement of isometric soleus muscle twitches**

Plan of instrument from above with the subject lying on left side and ball of foot resting on a 20 kg load cell embedded in a high inertia beam (weights not shown).

discharges of the soleus. The muscle contraction was captured with a 20 Kg load cell at the centre of a 1-metre-long high-inertia beam which was free to pivot about the axis of the ankle joint (figure 1).

Measurements were made after zeroing the instrument's goniometer to the right-angle position of the foot to the tibia. It was possible to stretch the calf muscle over a joint range of about 60° by applying dorsiflexing torques of 0 to 2·8 Nm with a printed motor coaxial with the beam and ankle joint. The recordings of four consecutive taps, with intervals to reset the system, were averaged and captured on a Medelec Sapphire 4ME EMG recorder (Medelec Ltd, Old Woking, Surrey, UK). Because of the high inertia of the beam, the system had rotated by no more than 1° by the time the contraction was recorded, making these measurements virtually isometric; any subsequent movement occurred after the contraction and relaxation phase and accounts for the force trace eventually falling below the baseline (figure 2).

The half-relaxation time (1/2 RT) corresponding to the inter-quartile relaxation time (time to fall from 75% to 25% of peak force) was measured in milliseconds (figure 2). Data was gathered at increasing joint torques to produce incremental changes in muscle length and since muscle length is linearly related to the joint position of the ankle,[13] specifying the joint angle specifies the muscle length. Sale and colleagues have shown that 60° of dorsiflexion from the resting joint angle correponds to about 6 cm of stretch for the adult soleus muscle.[13] Because of the known effects of temperature on muscle contractility, all the tests were done in a warm room. All legs were warm to the touch and none had evidence of erythrocyanosis or circulatory disturbance. The effects of surface electrical stimulation of the popliteal nerve with a current of 24 mA, pulse width 100 μs, in one of us (J-PL), elicited a muscle contraction comparable to the ankle jerk response (result not shown).

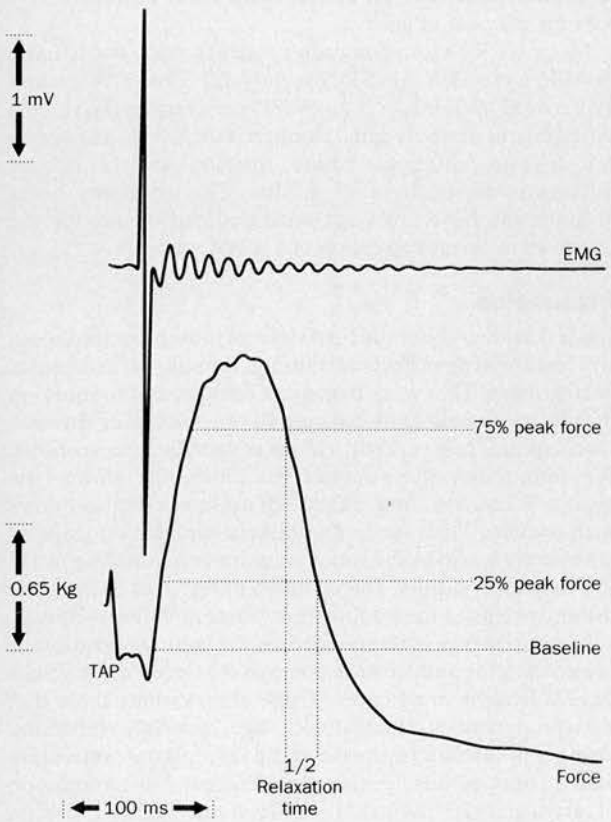

Figure 2: **Soleus muscle reflex EMG and muscle twitch force**

Force signal (lower trace) is obtained from the 20 kg load cell. Reflex electromyographic (EMG) discharge (upper trace) from surface electrodes. 19-year-old healthy male.

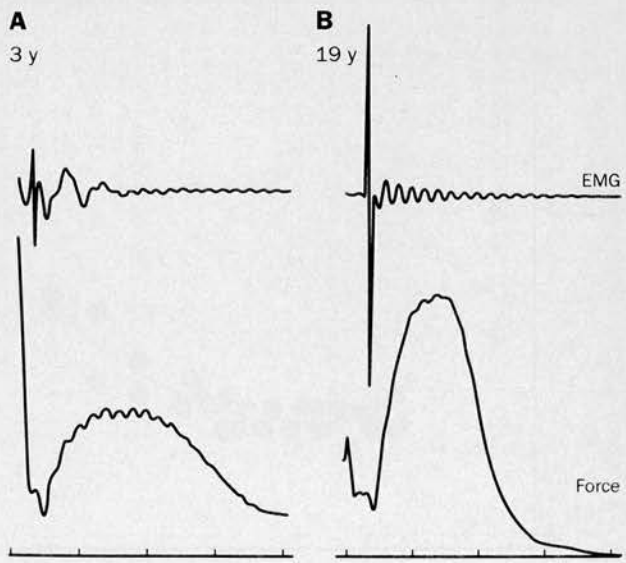

Figure 3: **Age and soleus-muscle relaxation time**

(A) Healthy 3-year-old male: PF = 0181 Kg, 1/2 RT = 75 ms. (B) healthy adult male: PF = 3 kg, 1/2 RT = 35 ms. Both contractions elicited at the neutral ankle joint position. Horizontal intervals = 100 ms, vertical intervals: for (A) force trace = 0·162 Kg, EMG = 250 μV; for (B) force trace = 0·65 Kg, EMG = 1 mV. (For abbreviations see text.)

Results

Figure 3 shows the difference between a healthy 3-year-old boy and a 19-year-old male: relaxation is prolonged in the child.

When the averaged isometric 1/2 RT elicited by tendon tapping is plotted against joint angle, it increases with increasing soleus-muscle length beyond the 0° (neutral) position irrespective of age as shown in figure 4 for a representative child and adult. The 1/2 RT against joint angle can be represented by a parabolic regression equation from which the value of the 1/2 RT at neutral ($x=0°$) is given by the constant a of the equation $y=a+bx+cx^2$. With exceptionally cooperative subjects, it is possible to make a large number of observations but in general fewer trials were done in younger children.

For valid comparisons between ages and the sexes, it is important to use values of 1/2 RT when the foot is at a right angle to the shaft of the leg, so that the 1/2 RT is compared at an equivalent muscle length for all subjects. When neutral 1/2 RTs, derived from quadratic regression equation for the combined data at all joint angles for the right and left soleus muscles of each child, are plotted against age for boys and girls aged 10 or under, a clear speeding up with age is evident as shown by the negative linear regression equation (figure 5), $r=0.71$. The 1/2 RT is roughly halved over the first decade of life from a mean of 90 ms at the age of 3 years (99% confidence intervals for the means of 1/2 RT = 70–110 ms) to about 40 ms at age 10. It can also be deduced from the slope coefficient of the regression equation that the expected 1/2 RT decreases by 6·61 ms per year of age.

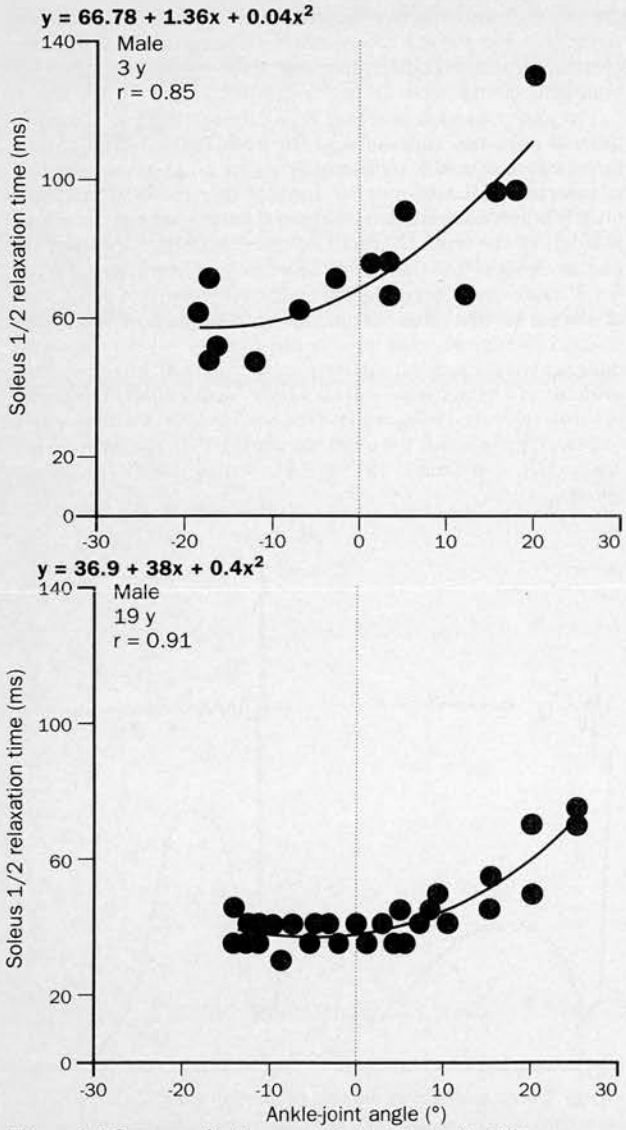

Figure 4: **Influence of soleus muscle length on the 1/2 relaxation time**

Irrespective of age or sex the values for the 1/2 RT against ankle joint position varies as $y=a+bx+cx^2$ (see results).

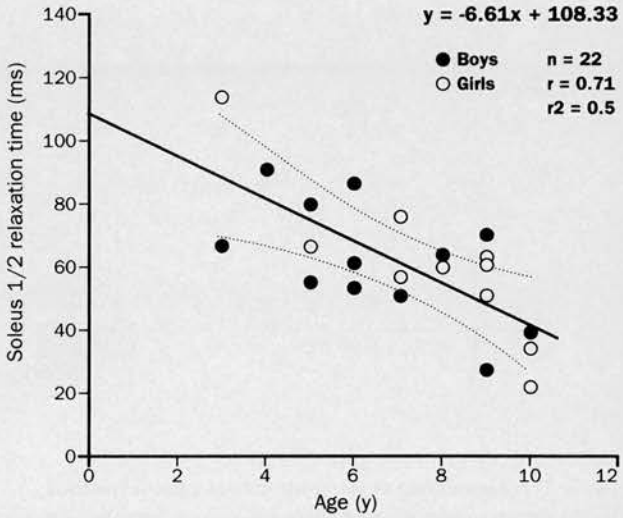

Figure 5: **Change in soleus half-relaxation time at neutral with age and sex**

The soleus 1/2 RT at neutral (0°) is plotted separately for 12 males and 10 females against age in years. The 1/2 RT is halved by age 10 in both sexes, $r=0.71$, $p=0.0002$ for the linear regression equation.

Mean 1/2 RTs for young adult males (n = 85) and females (n = 91) were 35·6 ms (SD 7·9, 99% CI 33·4–37·9 ms and 41·1 ms (SD 9·1, 99% CI 38·6–43·7 ms), respectively. This difference in means is small compared to the effect of age on 1/2 RT in children, whose muscles clearly behave differently from those of adults. The children, being prepubertal, have not been separated out by sex for the purposes of linear regression of 1/2 RT with age.

Discussion

Little is known about the behaviour of muscle in childhood, still less about the effects of training, trauma, brain damage, or treatment. This work began as a background to studying the effects of early brain damage on the muscles of children with cerebral palsy and the effects of specific interventions. We found that the younger the child, the slower the muscle-relaxation time. Although muscle relaxation slows with cooling,[14] this seems an unlikely explanation since all limbs were warm to the touch; to achieve a doubling in 1/2 RT in healthy adults, Davies and Young[14] had to immerse the entire legs of their subjects in water at 0° for 45 mins.

Measurements corresponded to the neutral position of the ankle joint and so were compared at equivalent soleus muscle lengths in all cases. These observations show that muscle dynamics change with age, possibly reflecting changes in calcium re-uptake of the sarcoplasmic reticulum which controls muscle relaxation. Postnatal differentiation of rabbit and rat slow muscles into fast muscles over the first three weeks of life has been attributed to changes in the macromolecular composition of muscle intracellular membranes, in particular mitochondrial glycerol-1-P dehydrogenase which is a specific mitochondrial membrane

enzyme related to the contractile characteristics of muscles and also to calcium-ATPase concentrations which are low at birth. In addition, enzyme systems characteristic of slow muscles such as cAMP-activated protein kinase abound at birth but eventually the whole mitochondrial population originally present in slow muscles is replaced.[15]

We believe the slower relaxation to be due to an intrinsic property of the muscle membranes either under the influence of a genetic developmental clock or capable of expression according to environmental stimuli such as the pattern of motor activity. Although we have confined our observations to the soleus muscle, observations of rapid alternating movements in muscles in the hand and forearm show a similar increase in speed with age (unpublished results).

Treatments for cerebral palsy remain empirical and most physicians, surgeons, and physiotherapists working with affected children would admit to an ad-hoc approach to the disordered motor system. Most systems of motor assessment remain descriptive and offer no means of predicting the likely physiological effects of a particular intervention. Preliminary results from chidren with congenital hemiplegia (in preparation) indicate that physical loading of the soleus muscle such as stretches and exercises induce slow-twitch characteristics whereas disuse, prolonged immobilisation, or unloading such as tendon releases produce fast-twitch characteristics which contribute to spasticity.

The deleterious morphological and physiological effects of muscle unloading by combinations of tenotomy and immobilisation have recently been reported[16] and such independent studies support the routine use of objective measurements of the contractile properties of muscles undergoing treatment not only to increase our understanding of human muscle physiology but to ensure better treatment for children with cerebral palsy.

We thank the children for their patience, adult controls for their participation, Dr R Baxendale of the Institute of Physiology, Glasgow University, and Dr Karen Kwong, Visiting Fellow from Hong Kong, for her assistance with some of the studies. Lesley Skeates-Bailey produced figure 1. The equipment costs were supported in part by the James and Grace Anderson Trust. JPL is supported by an Edinburgh University George Guthrie Research Fellowhip.

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Pulmonary immunopathology of sudden infant death syndrome

William J Howat, Isabella E Moore, Mary Judd, William R Roche

Summary

Sudden infant death syndrome (SIDS) is the most common cause of postneonatal mortality in the UK. Pathological investigations have shown evidence suggestive of respiratory obstruction with subsequent hypoxia leading to death.

We examined 48 infants who died of SIDS and 30 who died of other, non-pulmonary, causes and identified pulmonary eosinophil and neutrophil leucocytes, mast cells, and T and B lymphocytes by immunocytochemistry. Positively stained cells were counted in the parenchyma and around the bronchi without knowledge of the tissue source. The results showed three times more eosinophils in the lungs of infants who died of SIDS (27·61 *vs* 7·91 [99% CI 1·76–5·87] cells/mm² for parenchyma) accompanied by increased T lymphocytes and B lymphocytes. There were more peribronchial mast cells in the SIDS group (22·1 *vs* 14·7 [1·03–2·10] cells/mm²) and insignificant differences in neutrophils and parenchymal mast cells. There were significant associations between eosinophil, B lymphocyte, and T lymphocyte numbers.

These findings provide evidence for an abnormal T lymphocyte-mediated pulmonary inflammatory response in SIDS. Products of eosinophil degranulation can cause epithelial damage and pulmonary oedema, which could cause the respiratory obstruction and hypoxia associated with SIDS.

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Introduction

Although there has been a recent fall in the incidence of sudden infant death syndrome (SIDS), it remains the most common cause of postneonatal mortality.[1] Despite recent advances in understanding of child-care practices as a contributory factor to SIDS,[2] the underlying mechanisms and causes remain obscure.

Necropsy studies of SIDS have shown frothy, blood-tinged fluid around the nose and mouth in 50% of cases and intrathoracic subserosal petechial haemorrhages in 80%,[3] findings that have been interpreted as evidence of respiratory obstruction immediately before death. Furthermore, haemosiderin-laden macrophages in pulmonary tissue[4] and brainstem gliosis[5] are evidence of previous episodes of respiratory obstruction with hypoxia. Overall, these findings suggest that respiratory obstruction may be the final common pathway in SIDS.

SIDS is more prevalent in winter[6] in both the northern and southern hemispheres.[7] An inverse relationship has been described between incidence of SIDS and mean monthly temperature,[8] and a study of this association has indicated that a fall in temperature is followed approximately 6 days later by an increase in the incidence of SIDS.[9] Seasonal variation in SIDS has drawn attention to the role of respiratory viruses; several studies have shown a relationship between viral infections in the pre-school population and SIDS,[10,11] particularly for respiratory syncytial virus. Peak incidence of SIDS occurs when infants are particularly susceptible to infection due to waning maternally derived immunity.

We investigated the hypothesis that SIDS is an abnormal immune response in vulnerable infants to common respiratory pathogens by investigating inflammatory cell populations in SIDS and in infants dying from non-pulmonary causes.

University Pathology, University of Southampton, Southampton General Hospital, Southampton SO9 4XY, UK (W J Howat BSc, I E Moore MD, M Judd FIMLS, W R Roche MD)

Correspondence to: Dr William R Roche

Materials and methods

48 consecutive cases of SIDS from the Southampton General Hospital archives, 1985 to 1989, and 30 explained infant deaths from the Southampton and Oxford archives, 1974 to 1989, were examined. Necropsies had been done by paediatric pathologists within 72 hours of death. SIDS was defined according to established criteria,[12] and all cases had negative histological, microbiological, and virological investigations. The comparison group was infants who died of non-pulmonary causes in the first 18 months of life and in whom histology had revealed no pulmonary inflammation. Causes of death in this group were: trauma 9, congenital heart disease 8, necrotising enterocolitis 3, asphyxia 2, and 1 each of megacystis megacolon intestinal hypoperistalsis syndrome, haemorrhagic shock, meningitis, hydrocephalus, air embolus, post-operative respiratory depression, and intussusception with peritonitis.

Tissues had been fixed in neutral-buffered formol saline and embedded in paraffin. Lung-tissue sections were stained with haematoxylin and eosin and martius-scarlet-blue. Immunohistochemistry was done with streptavidin-biotin-peroxidase complexes[13] (Dako, High Wycombe, UK). Monoclonal antibodies to mast cell tryptase[14] (AA1, Dr A Walls, University of Southampton, UK), eosinophil cationic protein[15] (EG2, KabiPharmacia, Milton Keynes, UK), neutrophil elastase[16] (NP57, Dako), CD45RO[17] (UCHL1, Dr P Beverley, University College, London, UK), CD20[18] (L26, Dako), and a polyclonal CD3 antibody[19] (Dako) were used. Endogenous peroxidase was blocked with hydrogen peroxide in methanol and tissue-binding sites were blocked with avidin-biotin blocking kit (Vector Laboratories, Peterborough, UK) and incubation with tissue-culture medium containing 10% fetal calf serum.

Slides were coded and counted blind. Mast cells and eosinophils were counted in three areas each of bronchial wall and peribronchial tissue for each of three randomly chosen bronchi. T and B lymphocytes and neutrophils were counted in one area of wall and peribronchial tissue from 3 randomly selected bronchi; parenchymal cells were counted in 3 randomly selected areas of lung. Cell counts for the peripheral lung tissue were corrected for the percentage area occupied by alveolar walls, which were measured by a different observer (MJ) by automated image analysis.

When data were decoded and tested for skew, cell counts and age at death were found positively skewed. Cell counts were incremented by one and log-transformed, and age at death was log-transformed to normalise distributions. Transformed data were analysed by multivariate analysis with age at death and sex as covariates. Statistical significance was $p < 0.01$ after accounting for

THE MATURATION OF MOTOR DEXTERITY: OR WHY JOHNNY CAN'T GO ANY FASTER

J-P Lin
J K Brown
E G Walsh

Recent work on the contractile properties of the soleus muscle in healthy children suggests that muscles almost double in speed over the first 10 years of life, and that this change in the muscle properties is produced principally by a faster relaxation time (Lin *et al.* 1994). We investigated the speeds of alternating movements at the ankle at different ages, and correlated the findings with measured twitch parameters of the muscle. Speeds of alternating movements at the metacarpophalangeal (MCP) and wrist joints were likewise recorded in the same subjects to determine if muscle maturation significantly contributes to motor dexterity in childhood.

Subject and Method

Height, weight, fibula length, foot size and maximum calf diameter and circumference were obtained in 38 children and eight adults, and dexterity was measured in 13 of these children aged 3 to 11 years, all of whom had already undergone measurements of the soleus muscle twitch characteristics (Lin *et al.* 1994). Dexterity measures were obtained in 11 adults four of whom had undergone muscle twitch measurements. No muscle twitch data were available for the muscles acting at the MCP or wrist joints. All the children and adults were healthy volunteers, free of neurological disease. Subjects for this dexterity study were recruited when it became apparent that young children's muscles had slower relaxation times (Lin *et al.* 1994). Children undergoing puberty were excluded by applying an arbitrary cut-off point at 11 years of age. With parental consent (in the case of children) and full ethical approval from the Lothian Health Board Paediatric and Reproductive Medicine Ethics Committee, the speeds of alternating movements were measured at three joints.

The equipment consisted of a battery-operated timer capable of measuring the number of signals from a foot- or finger-switch over a 10s period. Measurements of alternating dorsiflexion and plantarflexion movements at the ankle were made with the subject seated comfortably and the hip, knee and ankle joints flexed to 90°. The heel was in contact with the ground throughout, and the subject was instructed to tap the foot as fast as possible without the heel losing contact with the ground (Fig. 1a). For the index finger, the speed of alternating flexion and extension at the metacarpophalangeal joint was measured with the index finger fully extended (Fig. 1b). The forearm, wrist joint and hand were immobilised palm downwards to ensure that only alternating movements at the MCP joint were recorded. In the case of the

alternating flexion/extension movements at the wrist, the forearm was held pronated on the table with movement restricted to the wrist joint, allowing the hand and the rigidly extended index finger to activate the switch (Fig. 1c). Three consecutive sets of measurements were obtained at each joint for the the right and left ankle, MCP and wrist joints, producing a total of 18 measurements per subject.

Dexterity data were expressed as the number of taps/second (Hz), and three data points were plotted for the right and left ankles, MCP and wrist joints for all 13 children and 11 adults, producing 6 frequency data points for each joint. Dexterity curves were obtained for these cross-sectional data by plotting tap frequency against age. Male and female subjects are distinguished by different symbols, but the regression curve applies to the combined male and female data.

Results

The ability to perform rapidly alternating movements requires appropriate agonist/antagonist muscle timing in a triphasic pattern (Fig. 2), and the speed of such movements is fastest with the ankle joint held at a neutral angle relative to the tibia (0° of dorsiflexion). As the speed of alternating movements increases, the amplitude of ankle joint displacement diminishes, because the acceleration required to move it increases with the square of the frequency of movement. If the subject attempts to tap too fast, a combination of electrical co-contraction and overlapping of the mechanical contractions between the preceding and immediate contractions prevails to bring movement to a standstill.

The maximum rate of tapping of the foot over a 10s period in 13 children and 11 adults was measured in hertz (Fig. 3). Tapping speed doubled between 3 and 10 years of age. The paediatric data were tightly clustered for the right and left ankles, and between children, with an almost linear increase in tap frequency over the first decade. The adult data were more scattered, though they were clearly faster than in the children under 10 years. Similar findings were obtained when the subjects were asked to tap rapidly at the

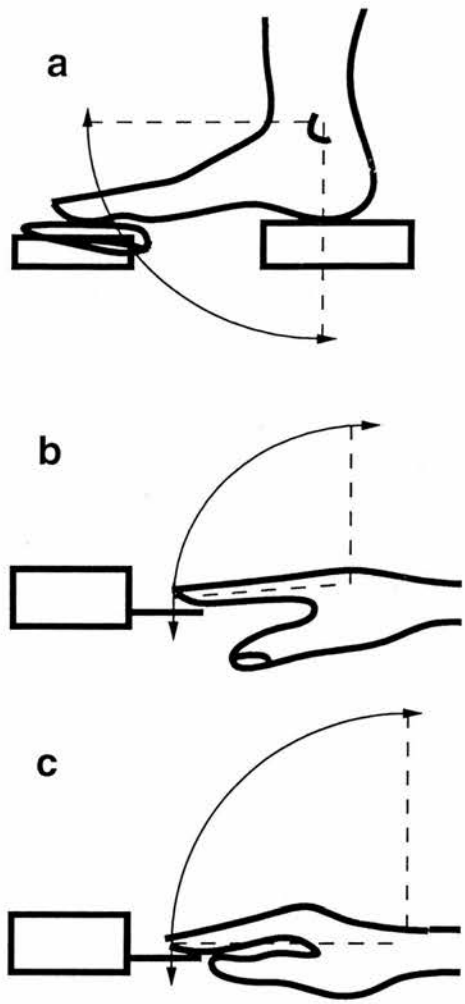

Fig. 1a: Axis and plane of rotation of foot about ankle joint, with foot switch activated by contact with ball of foot. ***b:*** *Index is flexed and extended about MCP joint, making contact with 'morse code' switch.* ***c:*** *Axis of rotation at the wrist joint: the hand is used as a 'door knocker' to activate morse code switch with tip of index finger.*

MCP joint of the index finger using long (forearm) extensors and flexors and the intrinsic hand muscles such as the first dorsal interosseus (FDI) and lumbrical muscles. When the three trials for each index were plotted against age, a steep increase in tapping frequency was seen and this also doubled over the first decade (Fig. 4). The fastest rates were obtained at the wrist for subjects of all ages but the effect of age was still striking (Fig. 5). On average, there was a

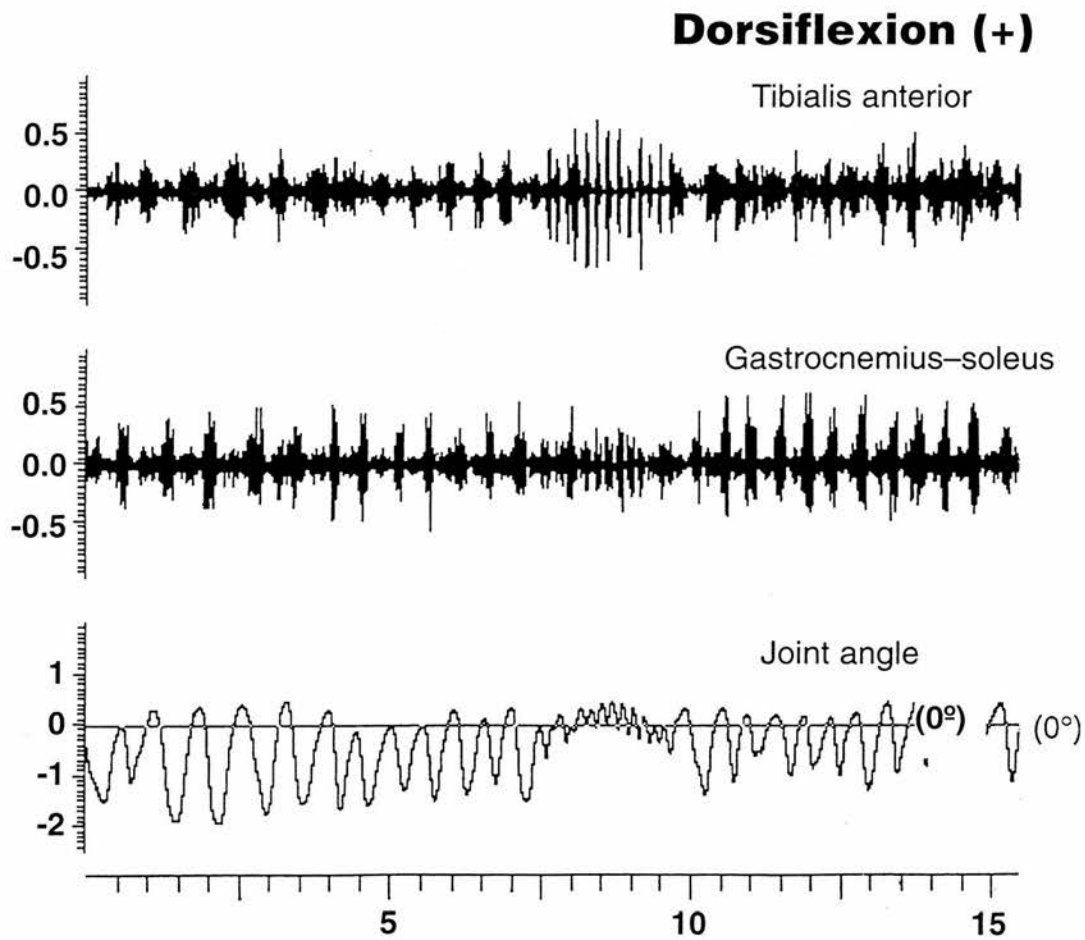

Fig. 2. *Optimal joint range for ankle dexterity – triphasic EMG response of tibialis anterior and gastrocnemius –soleus muscles. Joint angle for maximum speed of alternating movements is close to 0° of dorsiflexion,* i.e. *neutral position in which foot is at right angles to tibia. Amplitude of voluntary displacement varies inversely with square of frequency of movement. Vertical axis: 1 unit=30°*

0.75Hz difference between the tapping rate of the foot and that of the index, and an almost 2Hz difference between alternating movements at the ankle and at the wrist (Fig. 6), all three sets of data increasing in speed in parallel over the first decade and reaching a plateau by adulthood.

Correlation of speed of tapping at the ankle and soleus dynamics

The maximum speed of ankle tapping showed a high curvilinear dependency on the speed of the soleus half-relaxation time (Fig. 7) which was obtained in 13 children under 12 years and the four adults in whom soleus twitch times were measured, confirming that the muscle must have time to relax before a movement in the opposite direction is possible.

Limb length and limb inertia

The doubling in height with age is shown in Figure 8a and of fibula length and foot size in Figure 8b for 38 children and eight adults. Since limb inertia increases with the fifth power of limb length (Fig. 9), this doubling in the limb segment length corresponds to a 32-fold increase in the inertia of the limbs over time (Walsh and Wright 1987), despite which movements increased in speed during the first 10 years of life.

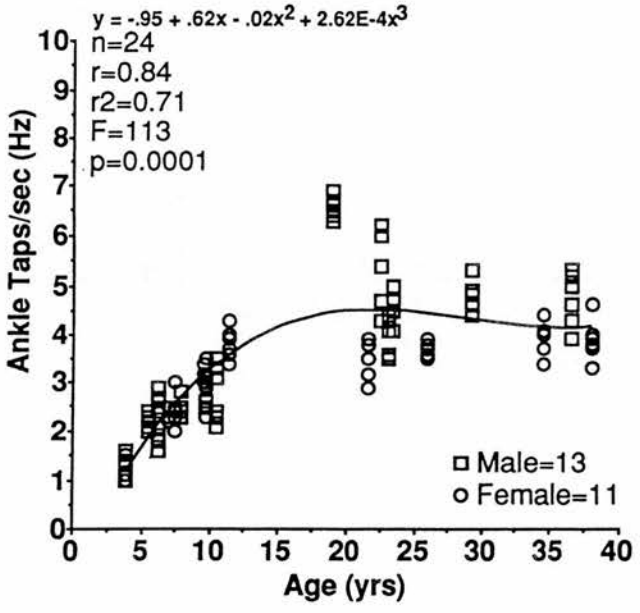

Fig. 3. Increase in dexterity at ankle joint with age. Three measurements of frequency of alternating dorsiflexion/plantarflexion at each ankle for each subject produces 6 data points per person, or 144 for 24 subjects. Most subjects produced tightly grouped values for each trial.

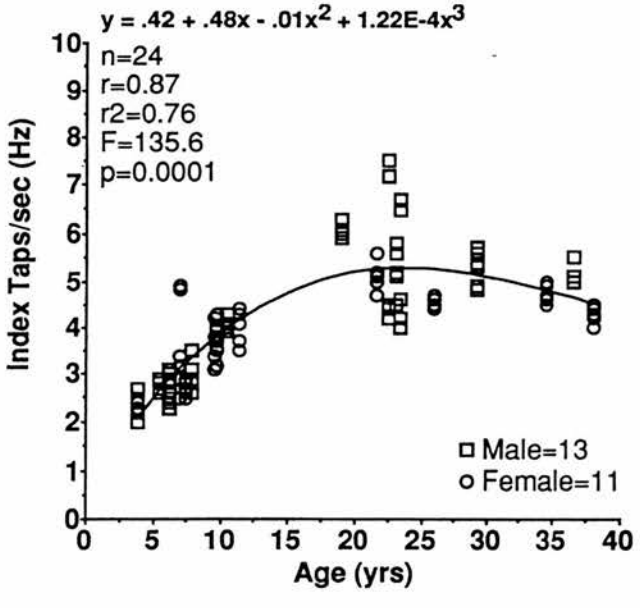

Fig. 4. Increase in frequencies of index tapping with age. (Flexion and extension at MCP joint)

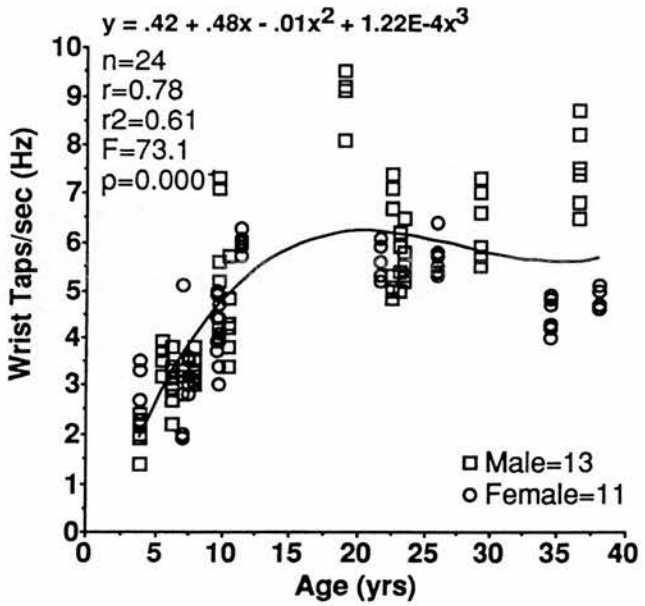

Fig. 5. Increase in frequencies of flexion and extension at wrist joint with age.

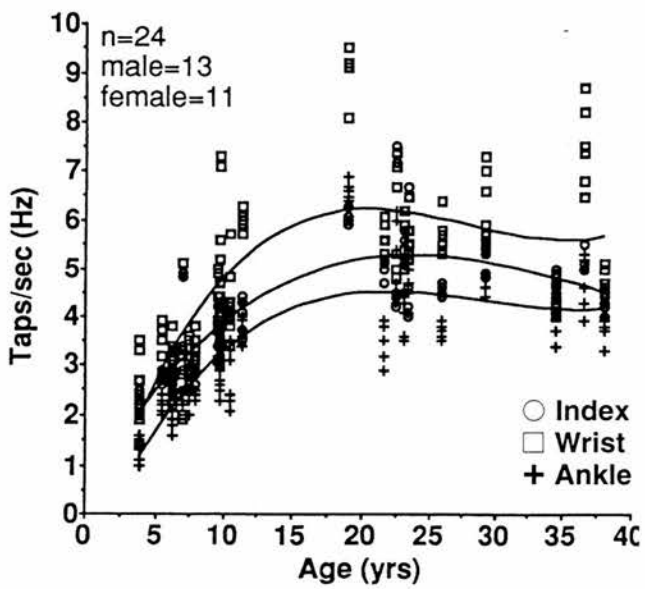

Fig. 6. Roughly parallel increase in dexterity at ankle, MCP and wrist joints with age.

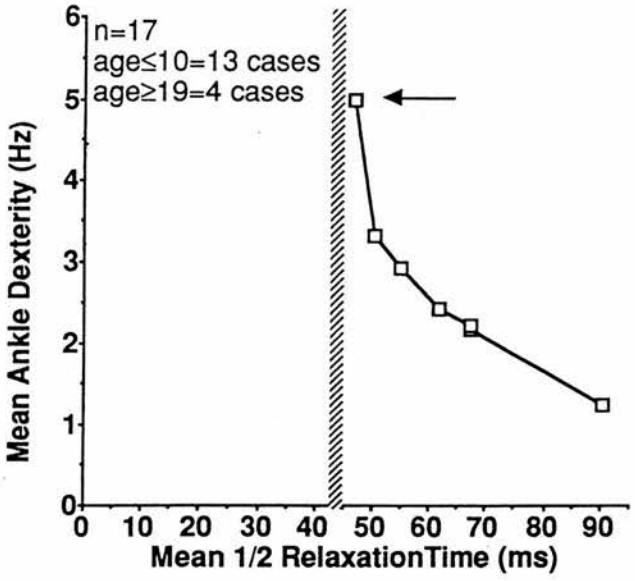

Fig. 7. Mean half-relaxation time of soleus muscle at neutral joint angle and ankle dexterity. Males and females combined: arrow indicates the four adult subjects. The ankle dexterity appears to vary inversely with the square of the half relaxation time.

Discussion

We have previously shown that the younger the child, the slower the muscle relaxation time, and we suggested that this must impose mechanical limits on the speed of motor activity (Lin *et al.* 1994). The present observations demonstrate the doubling in speed of alternating movements at the ankle, MCP and wrist joints between the ages of 3 and 10 years. This doubling in speed occurs despite a 32-fold increase in limb inertia produced by a doubling in limb length.

Several factors which contribute to the speed of alternating movements have been stressed. These include the functional joint range of the muscles involved and the timing of the agonist/antagonist EMG. Cerebral events and corticospinal and nerve conduction times control the timing of agonist/antagonist EMG activity, but the functional joint range is governed largely by peripheral factors: this implies that there is an optimal posture for fast movements at the ankle which corresponds to the neutral or right-angle position of the foot with the tibia. The reader can verify this fact by attempting rapid alternating plantarflexion/dorsiflexion movements at the ankle (1) in full plantarflexion, (2) in full dorsiflexion, and finally (3) at neutral. None of the extreme joint ranges feels either practical or comfortable: nor is it possible to perform forceful rapid movements in these positions. We have previously demonstrated that the relaxation time of the muscle is significantly prolonged in full dorsiflexion (Lin *et al,* 1994, Lin *et al,* forthcoming) when the soleus is stretched to its maximum length, and this in itself will reduce the speed at which an alternating task can be executed: an attempt at moving faster produces tetany of the muscles resulting in no net movement. In plantarflexion, however, the soleus is mechanically weak, so it is clear that anatomical considerations will affect muscle performance. At the optimal joint angle, the amplitude of the movements will be limited by the inertial characteristics of the foot and muscles and the relationship between acceleration and the frequency of oscillation which are governed by the equation:

$$\text{Acceleration} = (2\pi f)^2 A \sin 2\pi ft$$

where f=frequency, A=amplitude of displacement and t=time (Walsh 1992). Put simply, a doubling in the frequency of

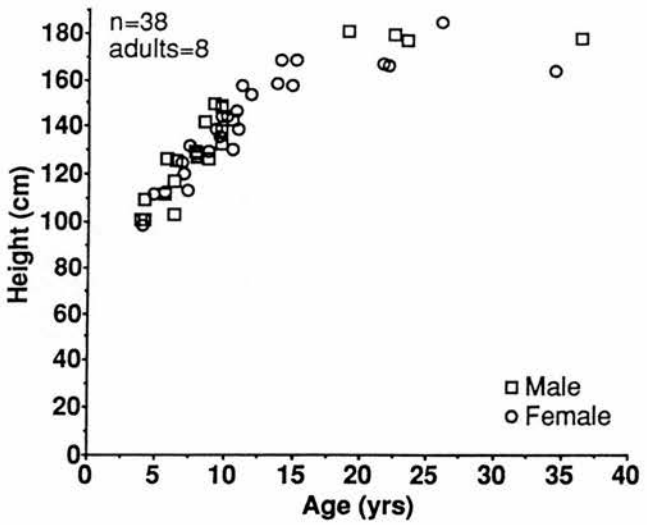

Fig. 8. Anthropometry. Height (top) and fibula length and foot size vs *age (bottom). Measurements from 38 healthy children and eight healthy adults.*

alternating movements would require a squaring of the acceleration involved. The body offsets this need for increased acceleration by adjusting the amplitude of movement. Slow movements requiring low accelerations can be performed at high amplitudes, whereas fast movements requiring large accelerations are performed at low amplitudes. Aside from these anatomical constraints, the speed of alternating movements is limited both by tetany and co-contraction of agonist/antagonist pairs.

Some of the mechanisms by which muscle dynamics may change with age, which have been previously reviewed (Lin *et al.* 1994), may reflect changes in the calcium re-uptake mechanism of the sarcoplasmic reticulum, which is known to control muscle relaxation. Animal studies have demonstrated postnatal differentiation of rabbit and rat slow

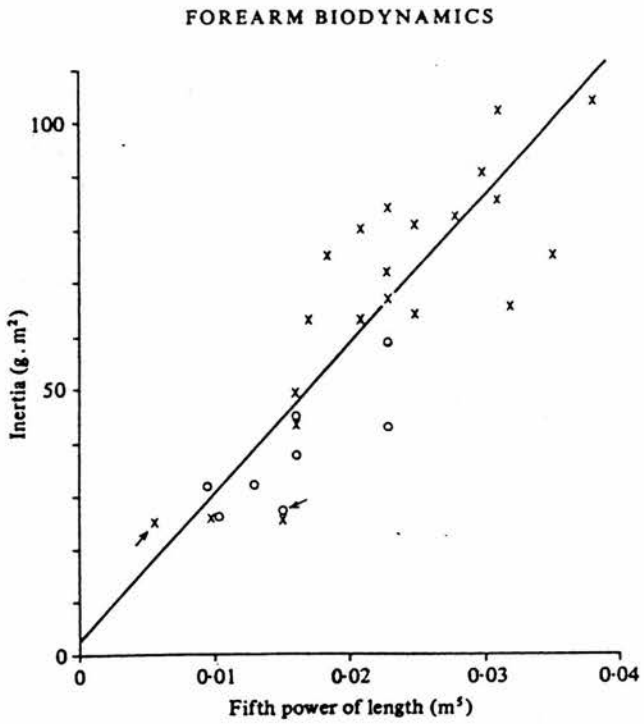

Fig. 9. Limb length and limb inertia (from Walsh 1987 with permission). Inertia varies with fifth power of limb length: i.e. *doubling in length results in a 32-fold increase in inertia.* ×, *male;* ○, *females. Data from the two children [in that study] indicated by arrows.* r=*0.84.*

muscles into fast muscles over the first month of life, which has been attributed to changes in the composition of muscle intracellular membrane enzymes such as mitochondrial glycerol-1-P dehydrogenase, a specific mitochondrial membrane enzyme related to the contractile characteristics of muscles, and also to changes in calcium-ATPase levels, which are low at birth. In addition, enzyme systems characteristic of slow muscles such as cAMP-activated protein kinase abound at birth, but the whole mitochondrial population originally present in slow muscles is eventually replaced with age (Margareth 1980).

Human motor development is particularly slow in comparison with that of other mammalian species. The change to an adult muscle phenotype of relaxation appears complete by about the age of 10, which also corresponds to the documented age at which a difference between fibre diameters in males and females occurs (Brooke and Engel 1969). In general, the faster the muscle phenotype, the more fatiguable it is. Scott and colleagues (1985) were unable to tire out the tibialis anterior muscles of healthy children with repetitive electrical stimulation, supporting our observations that children's muscles have a slow, oxidative, non- fatigueable phenotype to begin with. In a more recent study of 22 subjects aged 5 to 36 who had died accidentally, it was possible to demonstrate that muscle cross-sectional area in the vastus lateralis more than doubled, while there was also a decline of some 20% in the proportion of Type 1 (slow-oxidative) fibres between the ages of 5 and 20 years, suggesting that the muscles tend to speed up in the first 20 years of life (Lexel *et al.* 1992). Elder and Kakulas (1993) studied the contractile properties of the anterior and posterior calf muscles in 19 newborn infants and 35 children aged 5 to 16 months, and demonstrated a speeding up

of the half-relaxation time (1/2 RT) of the tibialis anterior muscle between 5 and 16 months of age although on average, the 1/2 RT of the plantarflexors seemed to slow by 15ms over a similar period. Despite this, there were some infants whose muscles remained persistently slow throughout the study period, and as the authors pointed out in a parallel histochemical study of 43 subjects (ranging from 22 weeks gestational age to 28 years) there was an overall preponderance of type 1 muscle fibres in children compared with adults in triceps brachii, vastus lateralis, biceps brachii and tibialis anterior muscles. The same data indicated that, as an infant becomes a child and then an adult, the muscles undergo a fast–slow–fast phenotype: *i.e.* the contractile properties are not fixed over time but may change adaptively in keeping with changes in motor development.

One intriguing question is the nature of the trigger for these phenotypic changes in muscle fibre type. It would seem logical that children are best suited to certain motor tasks, *e.g.* colouring in, singing, dancing, running, jumping and playing musical instruments (all of which require the ability to perform rapid alternating movements) when the muscles are ready. The findings also accord with the work of Denkla (1973), who documented an increase in the speed of repetitive thumb and finger oppositions or successive oppositions between thumb and all four fingers in turn. Our tasks were simpler in that they were purely repetitive, and it has been shown that repetitive alternating movements are accompanied by increased cerebral blood flow to the motor strip in the precentral gyrus but without corresponding activation of the supplementary motor area. Simple repetitive alternating flexion–extension motor tasks are accompanied by increased cerebral blood flow to the contralateral primary sensorimotor area but do not appear to involve other cerebral centres (Porter and Lemon 1993, describing the work of Roland and colleagues). Since corticospinal conduction times achieve a maximum rate at 18 months of age (Eyre 1991), the increase in speed of alternating movements cannot be attributed to spinal maturation. Although the peripheral nerves are known to increase in conduction velocity with age (Parano 1993), the conduction latencies involved actually take twice as long from birth to adulthood woing to an increase in nerve length (Mayer and Mosser, 1973) The candidate organ responsible for this maturation in dexterity is therefore muscle itself. Apart from being smaller and weaker with more compliant muscles, the young child is also intrinsically slow. For a further discussion of muscle strength and speed, see Lin (forthcoming).

The phenomenon of muscle speed should be borne in mind when looking at motor development in other muscle systems (for example the larynx, lips and tongue in slow or 'clumsy' speakers), and likewise when attempting to determine the effects of early brain or spinal-cord damage and subsequent treatments on the muscles of children.

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Authors' Appointments
*J-P Lin, MRCP, Senior Registrar in Paediatric Neurology, Great Ormond Street Hospital for Children NHS Trust, Great Ormond Street, London WC1N 3JH, UK.
JK Brown, FRCP (Edin), Consultant Neurologist, Royal Hospital for Sick Children, Edinburgh.
EG Walsh, FRCP (Edin), Department of Physiology, University of Edinburgh

**Correspondence to J-P Lin, Paediatric Neurologist, Gait and Movement Laboratory, Newcomen Centre, Guy's Hospital, St Thomas's Street, London SE1 9RJ.*

SUMMARY
The speed of alternating movements at the ankle, metacarpophalangeal and wrist joints in 11 healthy children and 13 adults doubled between age 3 and 11 years, despite a 32-fold increase in limb-segment inertia produced by the doubling in limb length over the same period. The data for the children showed little or no practice effect. The speeds for the adults, though faster than those

for the children, were more widely dispersed, indicating the possibility that training might increase the speed of the slowest adult. The findings are consistent with a previous report demonstrating a parallel increase in the speed of calf muscles over the first 10 years of life and it is inferred that the increase in dexterity at the wrist and metacarpophalangeal joints also depends on an increase in muscle speed with age. Muscle maturation may impose a rate-limiting envelope for all motor tasks which is particularly evident in rapidly alternating movements. These findings have implications for training in sport and music and for the understanding of motor delay, clumsiness and speech difficulties.

RÉSUMÉ

La maturation de l'habileté manuelle

La vitesse des mouvements alternés de la cheville, des métacarpophalangiennes et du poignet, notée chez 11 enfants en bonne santé et 13 adultes, doublait entre les âges de 3 et 11 ans, en dépit d'une progession de 32 fois de l'inertie du segment de membre produite par le doublement de la longueur du segment de membre durant cette période. Le recueil des données d'enfance révélait peu ou pas d'effet de pratique. Les vitesses pour les adultes, bein que plus rapides que pour les enfants, étaient plus largement dispersées. Les données sont en accord avec un article précédent démontrant une augmentation parallèle de la vitesse des muscles du mollet durant les dix premières années de vie et l'auteur en conclut que le gain de dextérité au poignet et aux métacarpophalangiennes dépend également d'un gain de vitesse musculaire. La maturation musculaire peut imposer une enveloppe limiatrice pour toutes les tâches motrices en rapport avec des mouvements alternatifs rapides. Ces données ont des implications pour l'entraînement du sport et de la musique, et pour la compréhension du retard moteur, de la maladresse et des difficultés de langage.

ZUSAMMENFASSUNG

Die Reifung der motorischen Geschicklichkeit

Bei 11 gesunden Kindern und 13 Erwachsenen verdoppelte sich die Geschwindigkeit alternierender Bewegungen in den Knöchel-, Fingergrund- und Handgelenken zwischen 3 und 11 Jahren, trotz einer 32-fachen Zunahme der Glied-Segment Trägheit durch Verdopplung der Gliedlänge im selben Zeitraum. Die kinlichen Daten zeigten einen geringen oder keinen praktischen Effeckt. Die Geschwindigkeiten bei den Erwachsenen, obwohl schneller als bei den Kindern, zeigten eine größere Streuung, was auf die Möglichkeit hinweist, daß durch Trining die Geschwindigkeit des langsamsten Erwachsenen erhöht werden könnte. Die Befunde stimmen mit einem früheren Bericht überein, in dem eine parallelle Geschwindigkeitszunahme in den Wadenmuskeln im Verlauf der ersten 10 Lebensjahre gezeigt wurde und man folgert daraus, daß die Zunahme der Geschicklichkeit in den Hand- und Fingergrundgelenken auch durch eine Zunahme der Muskelgeschwindikeit mit dem Alter bedingt ist. Die Muskelreifung kann einen die Geschwindigkeit begrenzenden Rahmen für alle motorischen Aufgaben vorgeben, was bei den schnell alternierenden Bewegungen besonders deutilich wird. Diese Befunde haben Rückwirkungen auf das Training im Sport und in der Musik und auf fas Verständnis von motorischen Entwicklungsverzögerungen, Ungeschicklichkeit und Sprachproblemen.

RESUMEN

Maduración de la destreza manual

La velocidad de hacer movimientos alternativos a nivel del tobillo, articulaciones metacarpofalángicas y muñeca, en 11 niños sanos y 13 adultos, se duplicó entre los 3 y 11 años de edad, a pesar de un aumento de 32 veces en la inercia del segmento de extremidad a consecuencia de duplicarse la longitud del miembro en el mismo periodo. Los datos de la infancia no mostraron ningun efecto o muy poco. Las velocidades en los adultos, aunque más rapidas que en los niños, se dispersaban más ampliamente, indicando que la posibilidad del aprendizaje podría aumentar la velocidad de los adultos más lentos. Los hallazgos están de acuerdo con publicaciones previas que demuestran un aumento paralelo en la velocidad de los músculos de la pierna más allá de los 10 años de edad y se infiere que el aumento en la destreza a nivel de la muñeca y las articulaciones metecarpofalángicas depende de un aumento en la velocidad muscular con la edad. La maduración muscular puede imponer una velocidad límite en todas las tareas musculares, que se pone especialmente en evidencia en los movimientos rápidos alternativos. Estos hallazgos tienen importancia para el entrenamiento en los deportes y en la música y para la comprensión del retraso motor, torpeza y dificultades en el lenguaje.

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Joint angle modulation of reflex neuromuscular output at the ankle in man

J.-P. Lin †, J.K. Brown ‡ and E.G Walsh *

‡*Royal Hospital for Sick Children, Edinburgh EH9 1LF,* **Department of Physiology, University of Edinburgh, Edinburgh EH8 9AG and* †*Guy's Hospital, London SE1 9RT*

With full ethical approval, non-invasive observations of the variation in reflex excitability and twitch characteristics of calf muscles with changes in joint angle were studied in nine healthy adult subjects (six males and three females) using a previously described high inertia mechanical filter to obtain isometric measurements (Walsh *et al.* 1993). Typical effects of passive stretch on reflex EMG (top trace) and reflex mechanogram (lower trace) are shown in Fig. 1 for one subject, representing four tap-triggered averages at increments from resting plantarflexion (−18 deg) through 0 deg, the neutral angle (hindfoot at right angles to the tibia) and dorsiflexion to +30 deg. The reflex EMG rises and falls as does the peak force, though the EMG begins to wane before the peak force. The twitch time rises progressively.

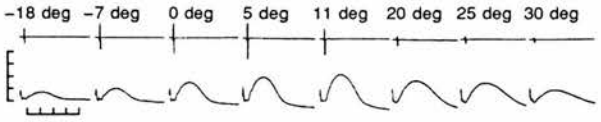

Fig. 1. Vertical interval = 1 mV for top traces (EMG) and 8·5 N for lower trace (force). Horizontal interval = 100 ms.

The tap-generated reflex EMG (Fig. 2*A*) is plotted against the joint angle for both limbs of all but one of the subjects, reaching a maximum about neutral and falling thereafter as does the peak twitch force (Fig. 2*B*) (normalized maxima for each limb separately).

Figure 2*C* shows the increase in twitch time with dorsiflexion, agreeing with data for direct electrical stimulation of the muscle by Sale *et al.* (1982); furthermore, passive muscle stretch modulates spinal motorneurone reflex output, the optimal joint angle for reflex muscular contractions occurring between −10 and +10 deg about the neutral joint angle.

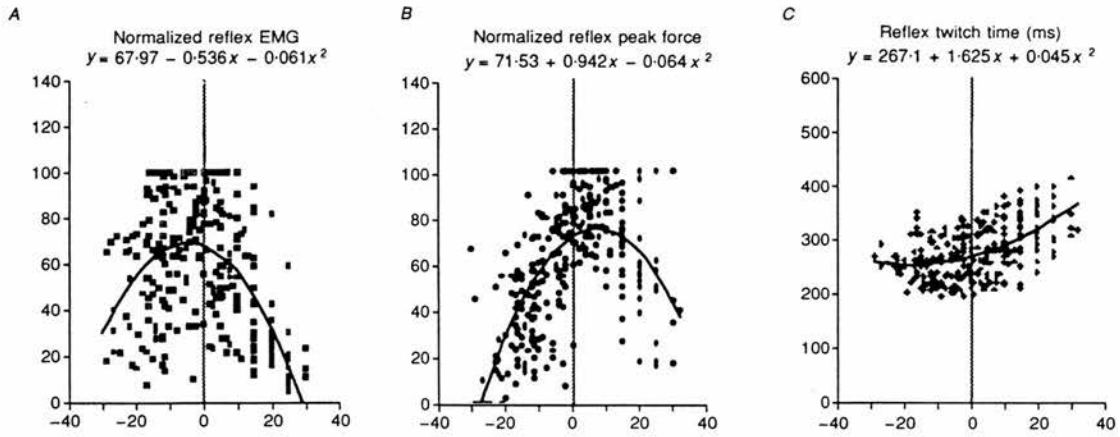

Fig. 2. *A*, EMG (%); *B*, peak force (%); *C*, twitch time (ms). Horizontal axis = degrees.

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Soleus muscle length, stretch reflex excitability, and the contractile properties of muscle in children and adults: a study of the functional joint angle

Jean-Pierre Lin* MRCP (UK), Consultant Paediatric Neurologist, 'One Small Step' Gait and Movement Laboratory and the Newcomen Centre, Guy's Hospital, Guy's and St Thomas's Hospital NHS Trust, St Thomas Street, London, UK.
J Keith Brown FRCPE, Consultant Paediatric Neurologist; **E Geoffrey Walsh** FRCPE: The Royal Hospital for Sick Children NHS Trust, Sciennes Road, Edinburgh, UK.

**Correspondence to first author.*

The influence of the joint angle on stretch reflex excitability of the soleus muscle at the ankle has been studied in 22 children aged 3.9 to 13.6 years and 9 adults aged 19 to 70 years. For all subjects, reflex EMG and mechanical twitch torque gain were trivial at resting plantar flexion. The reflex EMG gain reached a maximum between –15° and –10° of plantar flexion beyond the neutral angle, 0°, defined as the foot at right angle to the tibia, diminishing steeply with further dorsiflexion. The reflex mechanical gain rose to a peak between 0° and +10° of dorsiflexion beyond neutral, declining steeply thereafter. By contrast, axonally stimulated muscle twitch torque increased serially up to +30° dorsiflexion beyond neutral. For the soleus muscle, the optimal reflex neuromechanical angle lies approximately midway between the angle for optimal reflex EMG gain (in mild plantar flexion, at which the largest and strongest motor units can be activated) and the optimal muscle mechanical angle (at the extreme of soleus muscle dorsiflexion). These studies confirm that the excitability of the spinal alpha motor neuron pool in vivo is strongly influenced by muscle length and explain the variability in reflex excitability within and between subjects, if the joint angle is not controlled. They also indicate how posture influences movement, agreeing with the known function of the soleus muscle in the stance phase of gait and the modulation of motor unit recruitment during voluntary alternating movements at the ankle. Soleus muscle twitch characteristics show a fivefold to eightfold increase in peak force associated with a tenfold reduction in compliance in the first two decades of life and an apparent speeding up of twitch time in the first decade.

It has been known for more than 40 years that changes in joint angle alter reflex excitability (Magladery et al. 1951, Paillard 1959). Since then the mechanisms underlying modulation of the joint angle and its significance for the physiology and pathophysiology of motor function have become clearer. Herman (1970) indicated that the joint angle corresponding to maximum excitability of the gastrocnemius muscle was close to 30° of plantar flexion with the knee extended, whereas that for the soleus muscle was close to 90° (or the position at which the sole of the heel is at a right angle to the shaft of the tibia: the so-called neutral angle), indicating that for the soleus muscle, reflex excitability is reduced on either side of neutral, and for the gastrocnemius muscle, on either side of 30° of plantar flexion. Burke et al. (1971a) studied the effects of static stretching on the H (Hoffman) reflex (an electrically stimulated oligosynaptic reflex response) of the triceps muscle with the knee extended, and confirmed that stretches beyond neutral were reflex-inhibitory. They also confirmed, by means of ischaemic studies below the knee and above the ankle joint, that the mediators of the inhibitory influence resided in the length-sensing muscle spindles within the muscles rather than in the tension-sensing Golgi apparatus or cutaneous or joint capsule receptors (see discussion).

This effect of joint angle has been viewed as one of the confounding variables in assessing the monosynaptic tendon and H reflexes in pathological states, resulting in attempts to standardise methods of H reflex assessment by excluding the joint-angle effects to allow valid comparisons within and between subjects (Hugon 1973; see Method below). Robinson et al. (1982) demonstrated that stretching the triceps beyond neutral produced a 46.9% (± 19.2% SD) reduction in H reflex amplitude in all of the healthy subjects tested, whereas 5° to 15° of plantar flexion beyond neutral augmented the H reflex by 51.9% (± 42.1% SD) in 9 out of 10 subjects. Likewise, Tardieu and colleagues (1982) demonstrated an excitatory influence of the joint angle on the ankle plantar flexors when the triceps were stretched (dorsiflexed) from a position of relative plantar flexion through to neutral. The apparent continuum of reflex excitability has since been confirmed in a number of experimental conditions under which the muscles are active, such as treadmill walking and simple standing (Capaday and Stein 1986), in which maximal soleus H reflexes were obtained during the stance phase of gait, which is also the phase of gait during which the background electromyographic (EMG) activity is maximal. Similar results during the stance phase of treadmill walking were obtained by Crenna and Frigo (1987), who also looked at single-limb treadmill walking and stepping on the spot to separate out the possible effects of a bipedal motor task. A further report by Capaday and Stein (1987) indicated that the central alpha motor neuron drive such as that during running could override the influence of muscle length in reflex modulation, because although the EMG output was 2.4 times greater in running than in walking, the H reflex amplitude in running was either equivalent to or, often, lower than that obtained in walking. Gerilovsky et al. (1989) studied the effects of electrode montages (monopolar and bipolar), and of the positioning of surface electrodes along the midline back of the calf, on the H reflex amplitude. Although they found variations in the absolute H reflex amplitudes, depending on montage or electrode placement, the H/H_{max} ratio, or relative change in H reflex clearly altered with joint angle (muscle length), showing a 20 to 40% greater H reflex amplitude

with the foot plantar flexed to –30° (120° absolute joint angle) than at 0° (90° absolute joint angle) (with the knee extended). The greatest relative changes were captured by electrodes placed 1.5 cm and 3.0 cm below the insertion of the gastrocnemius muscle, whereas the greatest absolute changes were measured with electrodes placed 5.5 and 7.0 cm below the gastrocnemius insertion; however, with each and every electrode placement, a larger H reflex was obtained in –30° of plantar flexion compared with the neutral angle. Furthermore, identical relative recruitment curves were obtained irrespective of the use of a monopolar or bipolar electrode array with the calf muscle relaxed at an ankle joint angle of –10° (absolute joint angle 100°). Those authors stated: 'For a given electrode location the amplitudes of the monopolar motor unit (MU) action potentials increased with an increase in joint angle (i.e. decreased soleus length). The degree of the amplitude increase of the MU potentials was comparable to that obtained for monopolar H-potentials.'

More recently Brooke et al. (1995) have indicated that modulation of the H reflex, which is maximum in stance and inhibited in swing, may be influenced by movement at the hip and knee. Simonsen and colleagues (1995) have reported a task-dependent aspect to modulation of the H reflex, which is uniformly increased in stance in an uphill task compared with maximal at heel strike in association with co-contracting muscles acting across the ankle in a downhill task. Hultborn et al. (1996) have shown that the inhibitory influence of an initial stretching of static muscle may last up to 10 seconds or more after the muscle is restored to its original length. Further studies using transcortical magnetic stimulation (Hultborn et al. 1996) showed that alpha motor neuron excitability itself was not affected by muscle length, confirming the view that muscle length modulates presynaptic inputs to the motor neuron pool. In vitro studies by the same group showed that the motor neuron input resistance and membrane potentials were not influenced by a dorsiflexion conditioning stimulus, indicating that the principal effect of muscle stretch beyond neutral is to produce a moderately long-lasting homosynaptic postactivation depression.

Separate from these studies on modulation by the joint angle of the reflex and of recordable EMG potentials, the isometric mechanical torque of the muscles acting at the ankle joint in healthy adult subjects were extensively studied by Marsh et al. (1981) for the tibialis anterior and by Sale et al. (1982) for the plantar flexors. Electrical stimulation studies in vivo have shown that maximal torques are not obtained at the position of rest of the muscle; indeed, whether obtained as sustained maximal voluntary contractions or indirect electrical stimulation of the muscle by its nerve supply, the maximum torques of the tibialis anterior were obtained at about 10° of plantar flexion in the case of maximal voluntary contractions or in the case of tetanisation at frequencies of 20 to 40 Hz, whereas a 10-Hz frequency of electrical stimulation or single muscle twitches produced torque maxima at 30° of plantar flexion – i.e. in the almost fully elongated position of the muscle–tendon complex. Likewise, studies of the plantar flexors (Sale et al. 1982) indicated that, with the knee flexed at 90°, maxima of isometric torque were obtained at 20° of dorsiflexion beyond neutral for muscle twitches, tetanisation at 10 Hz, and the maximal voluntary contraction. The plantar flexor torque output is greater with the knee extended, i.e. when the gastrocnemius is stretched (see Silverskjöld 1923), although the influence of knee extension was not as marked as the authors had expected, the maximum influence of the gastrocnemius occurring between 25° and 10° of plantar flexion. The overall conclusion of the authors of the various studies on the dorsiflexors and plantar flexors at the ankle is that the maximum torques developed by these muscles occur when they are stretched, and that contrary to what is conventionally taught, the optimal angle is not the angle corresponding to the resting length of the muscle.

The present painless and non-invasive studies in adults and children look at the influence of static changes in the joint angle on mechanical reflex twitches obtained by tendon tapping at the ankle in terms of the reflex EMG, peak torques, and twitch times, with a view to correlating the electrophysiological and mechanical events. The study aims to bridge the gap in our understanding of the effect of joint angle on reflex electrical events and the ensuing reflex mechanical events, and to contrast this with the effect of joint angle on the axonally stimulated muscle. The findings, which have been briefly reported for healthy adults (Lin et al. 1996b), strongly support the concept of a continuum of reflex excitability, and of a functional joint range within which an optimal 'neuromechanical' joint angle can be specified. Furthermore, the optimal muscle length is different from the 'resting length'. This study is part of a broader investigation into the regulation of motor control (Lin et al. 1996a) and the influence of posture on dexterity (Lin 1997). The findings confirm the role played by peripheral factors in the subtle regulation of apparently 'centrally-defined' motor phenomena and provide predictive information on the likely behaviour of the motor system in health and disease for the clinician and therapist alike. These studies complement our previous clinical analysis of the pathophysiology of equinus in childhood hemiplegia (Lin and Brown 1992) and the assessment of spasticity in hemiplegic cerebral palsy (Lin et al. 1994a,b).

Method

With full approval of the Lothian Health Board Paediatric and Reproductive Medicine Ethics Committee, 31 healthy subjects were studied: 9 adults aged 19 to 70 years and 22 children aged 3.9 to 13.6 years. All the studies took place in the same warm room under similar conditions. Parents gave informed consent and were present throughout for all children studied. A typical study lasted an hour for two limbs, with time to rest between limbs.

The apparatus and experimental conditions for measuring a virtually isometric soleus muscle twitch after a tendon tap have been previously reported (Walsh 1992; Walsh et al. 1993; Lin et al. 1994c, 1996b). Measurements were made with the subjects lying comfortably on the left side with the knee flexed to 90° (Figs. 1 and 2) to eliminate the effects of the gastrocnemius muscle, which crosses both the knee and ankle joints (see Silverskjöld 1923). The ankle joint is coaxial with the axis of a horizontal beam which is free to rotate but is heavily weighted at each end to increase the beam inertia, thus acting as a mechanical filter. The angular position of the ankle joint is monitored by a potentiometer zeroed to a 90° angle between the sole of the heel and the shaft of the tibia. The position of the ankle joint, and hence the length of the soleus muscle, was varied by means of a printed electrical motor coaxial with the beam capable of applying a graded torque up to a maximum of 2.8 Nm (Fig. 2), so that each incremental joint angle was

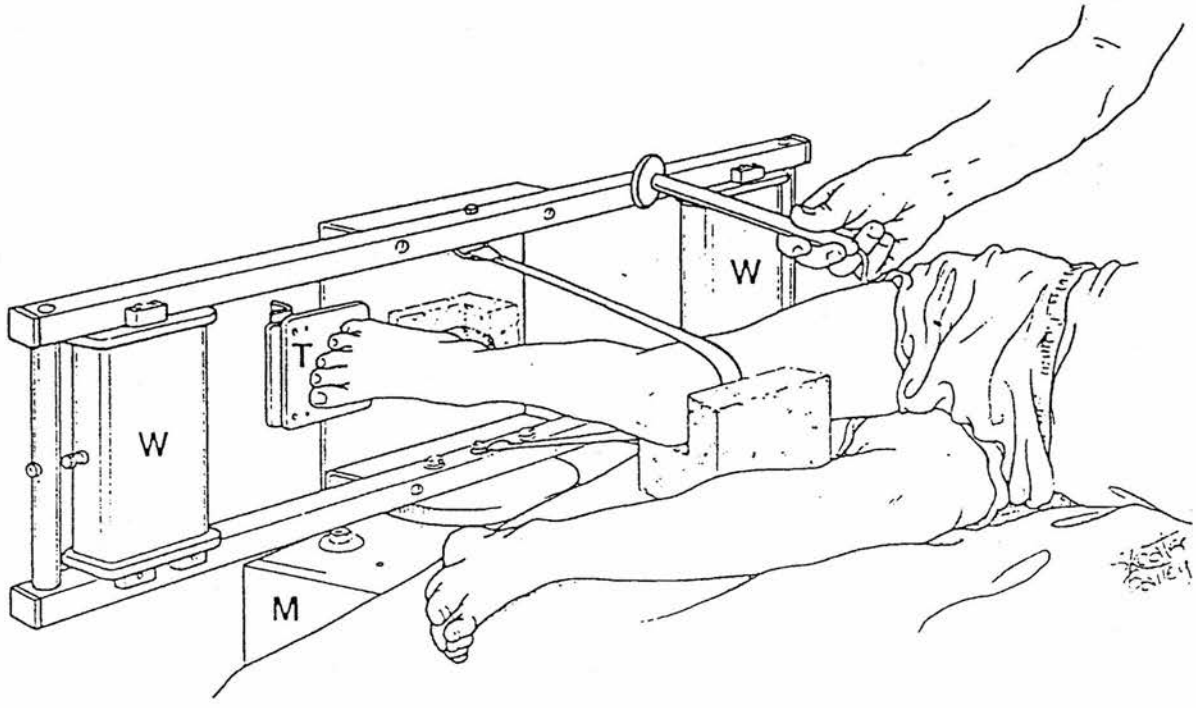

Figure 1: *Artist's impression of apparatus and arrangements for measuring near-isometric soleus muscle reflex twitches. Note heavily weighted ends (W) of beam to increase inertia, force plate (T) measuring tension generated by calf contractions, and coaxial printed electrical motor (M) to rotate beam. Reproduced by permission from Walsh (1992). Drawing by Lesley Skeates-Bailey.*

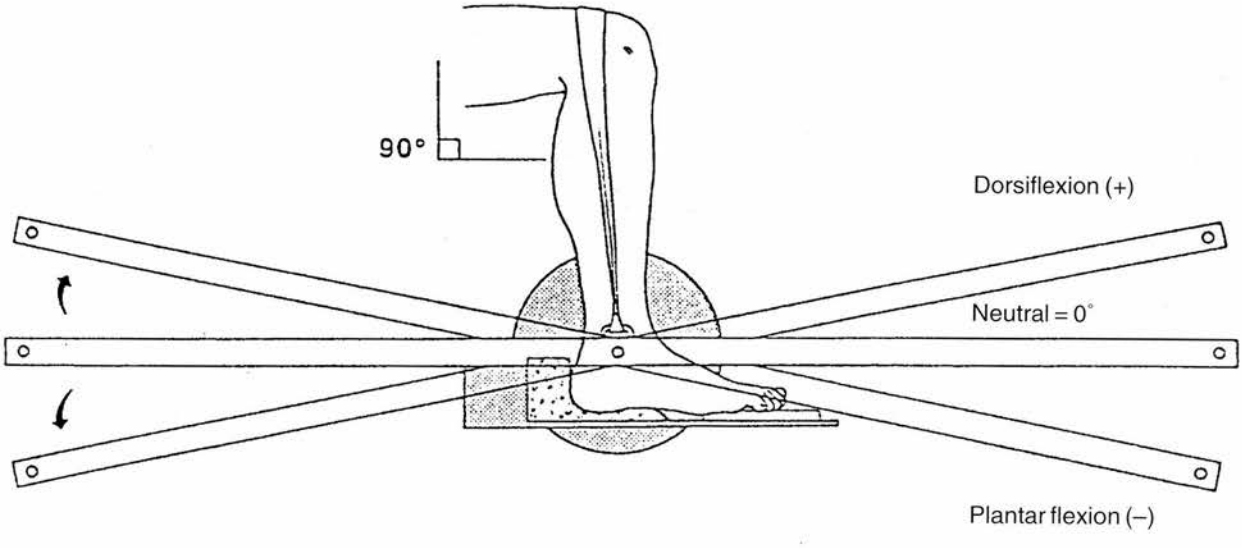

Figure 2: *Apparatus seen from above. Subject lies in left decubitus position to eliminate influence of gravity (see Fig. 1). Hip and knee are flexed at 90° to eliminate influence of gastrocnemius muscle, and ankle joint is coaxial with high-inertia beam, which is free to rotate. A printed electric motor, also coaxial with beam and ankle joint, applies incremental torques to dorsiflex foot from natural resting plantar flexion (negative joint angles) through neutral (0°, with foot at right angles to shaft of tibia) into dorsiflexion (positive joint angles). Ball of foot rests on 20-kg strain gauge plate to record mechanical twitch generated by a tap to Achilles tendon as shown in Figure 1. Surface reflex EMG is recorded by disposable electrodes placed in midline posteriorly at position of maximum calf diameter for active electrode and just above point of insertion of muscle into tendon for reference electrode. Averages of responses to four consecutive taps were taken at each increment of dorsiflexion maintained by electric torque motor. Drawing by Lesley Skeates-Bailey.*

achieved at a known torque. The plantar flexor force was sensed by a 20-kg-load cell embedded in the beam and in contact with the ball of the foot, and the results were converted to newtons (N), the raw data being expressed in newtons and the normalised data for each limb in units of percentage torque (% torque). One of the advantages of the beam method was that it obviated the need to constantly readjust the baseline force output signal with each new joint angle (see Walsh et al. 1993). The surface EMG was recorded from a bipolar arrangement of disposable silver–silver chloride electrodes (2.2 × 2.2 cm silver 'Mactrode', Marquette Electronics, USA) with the active electrode placed in the midline posteriorly at the position of maximum calf diameter, the reference electrode placed distally just above the insertion of the muscle into the Achilles tendon, and the earth electrode on the kneecap for each leg. This positioning of electrodes was chosen to simplify electrode placement in subjects of various sizes, whose ages ranged from 3 to 70 years. The EMG signal was amplified, filtered at 20 Hz to 10 kHz, and stored on a computer disk, along with the force and potentiometer signals, using a Medelec Sapphire 4ME recorder (Medelec Ltd, Old Woking, Surrey, UK).

Ankle reflexes were obtained in the resting muscle by manually tapping the Achilles tendon, using a tendon hammer with an electrical switch in the head capable of triggering the recorder screen. In all cases, four consecutive taps were delivered, with time between them to allow for reflex contraction and relaxation of the muscle. The results are averages, for the reflex EMG and reflex muscle twitch, of the responses to four taps at each given increment of joint position. When possible, measurements began at the resting joint angle, which was always in plantar flexion (equinus) relative to the neutral angle; however, in some individuals no reflex was obtainable until the soleus muscle had been stretched by applying a dorsiflexing torque with the motor. Throughout this study, 'negative angles' refers to plantar flexion and 'positive angles' to dorsiflexion beyond neutral (foot at right angle to tibia); neutral is referred to as the 0° angle. For each limb, reflex twitches were studied at small, sustained incremental torques of dorsiflexion from resting plantar flexion, through the neutral joint angle into maximum dorsiflexion. Some limbs were studied twice to confirm reproducibility and reliability data.

The characteristics of the reflex EMG and muscle twitch after a tendon tap are shown in Figure 3a, which depicts the averaged responses to four consecutive tendon taps. The mechanical tap, seen as a brief stimulus artefact on the force trace (N), causes a brief but large-amplitude EMG discharge (mV) some 12 to 35 ms later, depending on the subject's age and the length of the limb. This electrical discharge in the muscle is followed by a slowly rising mechanical contraction, reaches a peak, and then relaxes. The sequence *tendon tap – reflex EMG discharge – muscle twitch* is referred to in the text as 'neuromechanical coupling'. In a few adult cases, a fixed electrical current stimulus was applied percutaneously to the tibial nerve in the popliteal fossa to stimulate the soleus muscle via the motor axons, producing an EMG M response within a few milliseconds of stimulation, at various joint angles (Fig. 3b: averaged responses to four consecutive electrical stimuli). By varying the electrical stimulus current (50 to 100 mA, pulse width 100 μs), the spinal alpha motor neurons could be reflexly stimulated, producing the EMG H response some 30 to 35 ms later (Fig. 3b). This latency is the time taken for the electrical impulse to travel from the point of stimulation up the Ia afferent (large-diameter

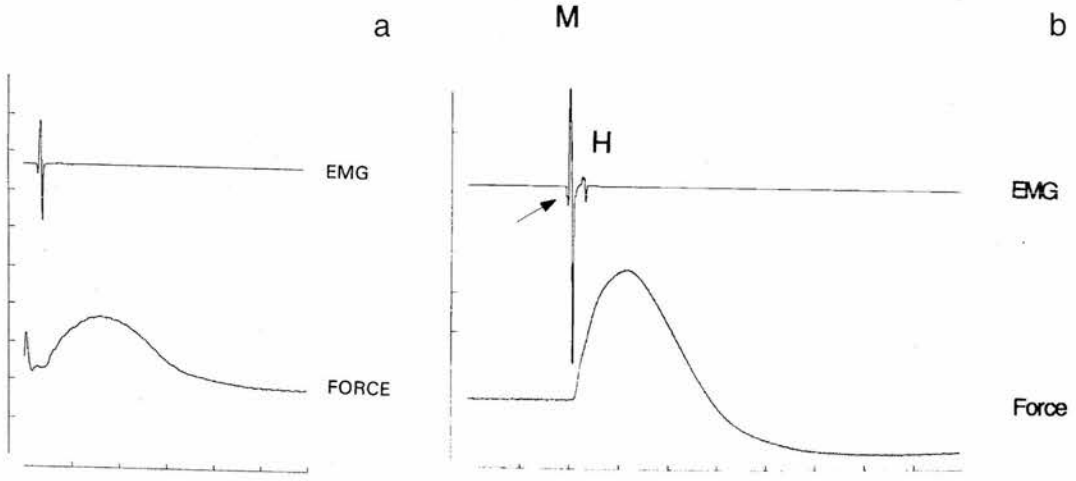

Figure 3: *Soleus muscle tendon tap reflex and direct axonal stimulation.* (a) *Tap force artefact (N) is seen as a brief deflection on force trace, followed 30–35 ms later by high-amplitude reflex soleus EMG discharge (mV) on EMG trace and is followed by slowly rising and falling mechanical twitch force (N). Tracings represent averages of responses to four consecutive taps, to compensate for variation in tap force between blows.* (b) *Stimuli consisted of a percutaneous fixed current of 56 mA with a pulse width of 100 μs, producing four consecutive stimulus-triggered twitches, which are represented as an average. Electrical stimulus (brief downward deflection) is on EMG trace and virtually coincides with brief, large-amplitude EMG discharge, called M response. This is followed some 30–35 ms later by onset of a brief, small-amplitude Hoffman* (H) *reflex. Note similarity in M and H response waveforms. Irrespective of stimulus mode, mechanical twitch force (N) rises and falls slowly on force trace. Twitches in a and b are similar. Horizontal intervals, 100 ms; vertical intervals, 12.8 N for force trace and 2 mV for EMG trace. M, direct motor response; H, Hoffman reflex. Male, aged 31 years, right soleus.*

sensory) fibres to the spinal cord, depolarise the spinal alpha motor neurons, travel back down the motor axons, and depolarise the muscle. This test was done to compare the effects of the joint angle on reflexly stimulated soleus muscle output (via the spinal cord) with that produced by directly stimulating the axons supplying soleus muscle, since direct stimulation bypasses the muscle spindle and the spinal cord apparatus. Because Hugon (1973) reported that the gastrocnemius muscle H reflex cannot be elicited when that muscle is at rest, all the H reflexes elicited in these studies are soleus reflexes. Hugon (1973) also stated that if the M and the H responses had the same electrical waveforms, they were likely to originate from the same muscle – in the present case, the soleus. In contrast to Figure 3a, Figure 3b shows three events on the EMG trace: i) an electrical stimulus artefact (arrowed small downward deflection), ii) a large-amplitude EMG discharge (M response) within a few milliseconds of the onset of the percutaneous electrical stimulus to motor axons of the tibial nerve, and iii) a small H reflex about 30 ms after the stimulus artefact. The force trace in Figure 3b shows no tendon tap artefact preceding the twitch, which is produced by the M response.

Clinical measurements included age (y), sex, weight (kg), height (cm), fibula length (cm), foot length (cm), maximum calf diameter (cm), maximum calf circumference (cm), resting joint angle (°), bias torque (N m), bias joint angle (°), and angular displacement (°); the soleus muscle compliance was obtained by dividing the angular displacement by the applied torque and was expressed in degrees/newton metre (°/N m). The neurophysiological measurements included: tendon tap force (N) (or, when normalised, the tendon % tap torque), reflex soleus muscle peak-to-peak EMG amplitude in millivolts (mV) or normalised EMG (% EMG), absolute peak force in newtons (N) or units of torque (% torque), half contraction time (interquartile contraction time, ½CT, ms), half relaxation time (the interquartile relaxation time, ½RT, ms), and total twitch time in milliseconds (ms).

ANALYSIS

The means and standard deviations of the applied bias torque (N m), measured calf compliance (°/N m), absolute reflex peak force (N), and reflex twitch time (ms) are plotted against age for the whole group. The tendon tap torques for each limb are normalised against the maximum value obtained to give % tap torque, which is plotted against the joint angle for each subject. The reflex EMG gain, defined as the ratio of normalised EMG to normalised tap torque (% reflex EMG/% tap torque), is plotted against the joint angle to examine the change in reflex gain against joint angle for all ages. Similarly, the reflex mechanical gain, defined as the ratio of normalised peak reflex torque to normalised tap torque (% reflex torque/% tap torque), is plotted against joint angle. The absolute twitch times are plotted against joint angle.

Results

EFFECTS OF AGE

The effect of age on the biomechanical and contractile properties of muscle is demonstrated for all 31 subjects in Figure 4, which depicts the mean and standard deviation across the joint range for a) applied bias torque (N m), b) soleus muscle compliance (°/N m; gastrocnemius excluded because the knee was flexed to 90°), c) reflex peak force (N), and d) twitch time (ms). The applied bias torque used to produce increments of

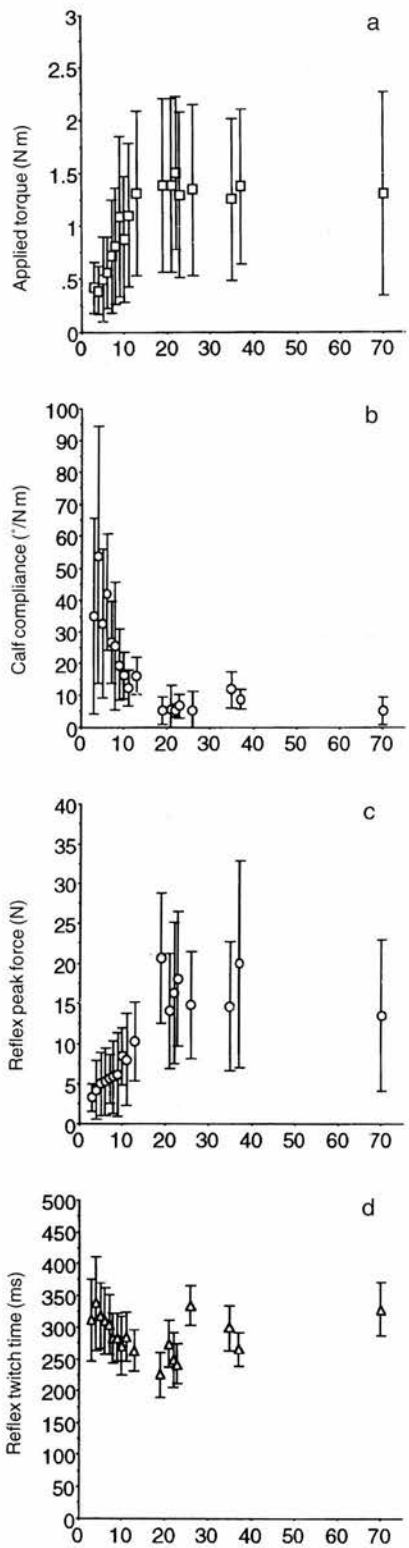

Figure 4: *Changes in characteristics of soleus muscle with age. Mean and standard deviations for 31 healthy subjects aged 3.9–70 years, some subjects having the same age. (a) Mean applied dorsiflexing torque (N m) rises as (b) mean soleus muscle compliance (°/N m) diminishes in first decade of life, while (c) reflex peak twitch force (N) increases and (d) mean reflex twitch time (ms) falls over same period.*

dorsiflexion clearly increases with age: the older the subject, the greater the dorsiflexing torque required to produce equivalent angular displacements. There is a dramatic, tenfold decline in mean calf compliance (excluding gastrocnemius) in the first two decades of life, indicating a real change in the biomechanical properties of the skeletal muscle fibres and connective tissue with age, which in turn must affect muscle performance. The mean peak reflex force increases fivefold to eightfold with age, whereas the mean twitch time diminishes by 30%, from 325 ms to 225 ms between 3 and 19 years, slowing again thereafter for the seven subjects over 19 years. The corresponding mean twitch frequency (inverse twitch time, not shown) increases from 3 Hz to 4.5 Hz.

EFFECTS OF THE JOINT ANGLE (SINGLE SUBJECT)

When the ankle reflex is elicited at sustained increments of dorsiflexion from the resting plantar flexor angle, through neutral, to maximum dorsiflexion, four phenomena are evident (Figs. 5 and 6a,b,c,d).

i) EMG gain

The reflex EMG gain (normalised reflex EMG/normalised tap torque ratio; Fig. 6b) reaches a peak and doubles close to –5°, diminishing with further dorsiflexion beyond neutral, until at +30° the reflex gain is a fraction of the peak and resting values. No reflexes were obtainable in plantar flexion beyond the resting angle. The EMG gain diminishes from +10° to +30° despite the fact that the tap torque (Fig. 6a) remains constant. The joint angle (muscle length) appears to modulate the recruit-ability of the motor neuron pool by initially *facilitating* and then *inhibiting* the reflex discharge, overriding the strength of the tap stimulus.

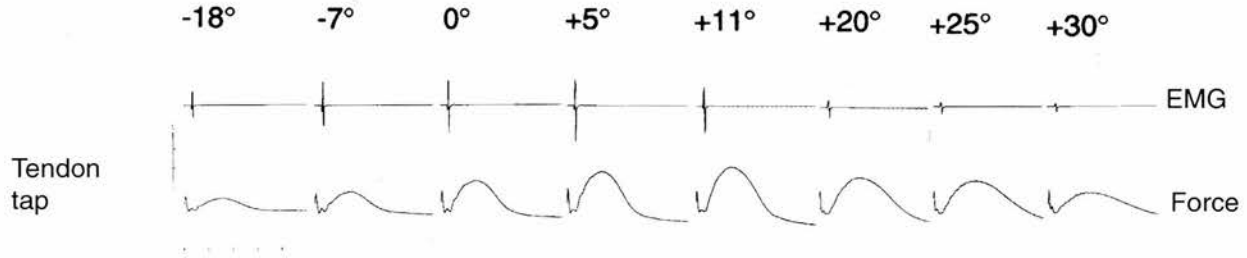

Figure 5: *Effect of joint angle on reflex EMG, twitch force, and twitch time. Reflex EMG (*top trace*) and peak force (*bottom trace*) become maximal close to neutral but wane rapidly in extremes of either plantar flexion or dorsiflexion. Muscle twitch time increases serially with each increment of dorsiflexion beyond neutral (see Fig. 6). Healthy adult, right soleus; each trace represents an averaged response to four taps. Horizontal intervals, 100 ms; vertical intervals, 12.8 N for force trace and 2 mV for EMG trace.*

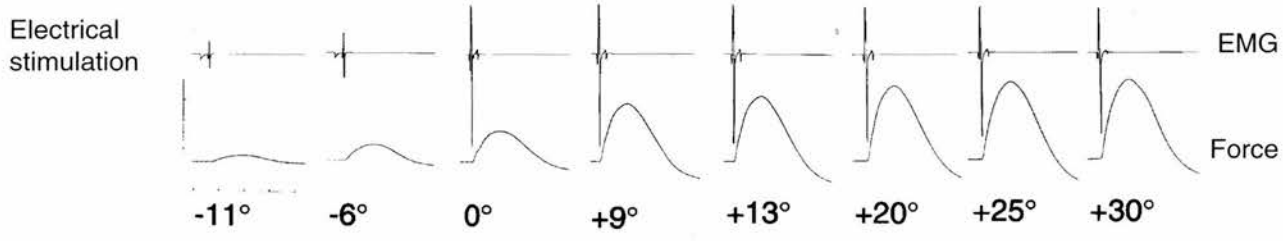

Figure 7: *Percutaneous axonal electrical stimulation of soleus muscle. Same subject, experimental session, and electrode positions as mechanical reflex studies in Figures 5 and 6. Each trace represents an average of responses to four electrical stimuli. At –11° and –6°, percutaneous axonal stimulation of posterior tibial nerve produces a small electrical stimulus artefact (downward deflection) on EMG trace followed by an H reflex EMG discharge some 30–35 ms later. From 0° onwards, same electrical stimulus produces a large direct EMG discharge (M response) which occurs within a few milliseconds of electrical stimulus and is then followed by a small H reflex some 30 ms later, reaching maximal amplitude at –6° (see Figure 3 for clarification of M and H responses). EMG waveforms of H and M responses are similar to tap-induced reflex EMG discharges of Figure 5, indicating a common soleus origin (Hugon 1973). M response varies little from 0° to +30°, but peak twitch force rises serially to a maximum at +30° (optimal mechanical angle). By contrast H reflex diminishes at dorsiflexion beyond neutral. Horizontal intervals, 100 ms; vertical intervals, 12.8 N for force trace and 2 mV for EMG trace.*

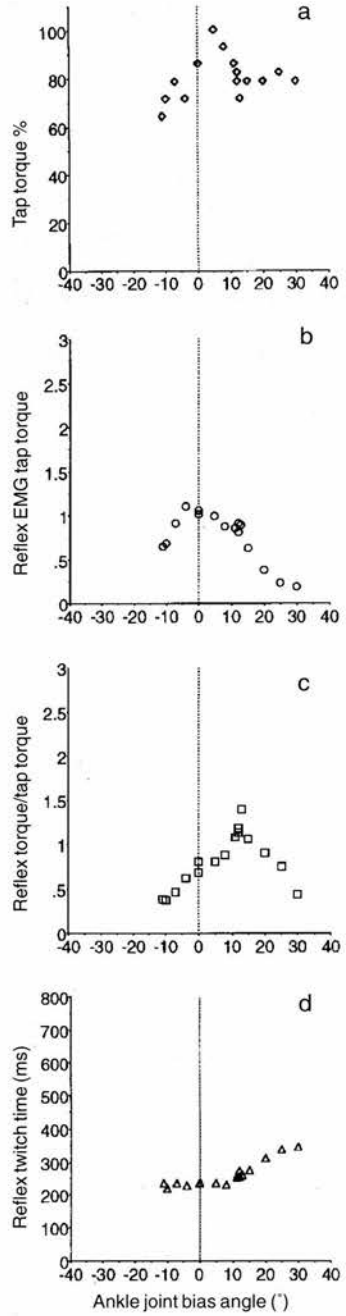

Figure 6: *Reflex EMG and mechanical gain with changing joint angle. Graphical representation of data from Figure 5. (a) Normalised tap torque (% tap torque) across joint range. (b) Change in reflex EMG gain (% reflex EMG/% tap torque) reaches a peak at −5°, which is 63% greater than resting gain and 450% greater than gain at +30°. (c) Reflex mechanical gain (% peak twitch torque/% tap torque) against joint angle reaches a peak at +10° to +15° of dorsiflexion beyond neutral. There is a 265% difference between resting and peak mechanical gain and 221% fall in mechanical gain at +30°. Reflex mechanical output is maximal at midjoint range, more than 20° dorsiflexed beyond resting angle. (d) Muscle twitch time (ms) remains constant at 225 ms from −12° (resting angle) to +10°, increasing steeply to 375 ms at +30° of dorsiflexion beyond neutral. Right leg. 31-year-old adult male: each point represents an averaged response to four consecutive taps. 0°, neutral angle: vertical dotted line in all graphs.*

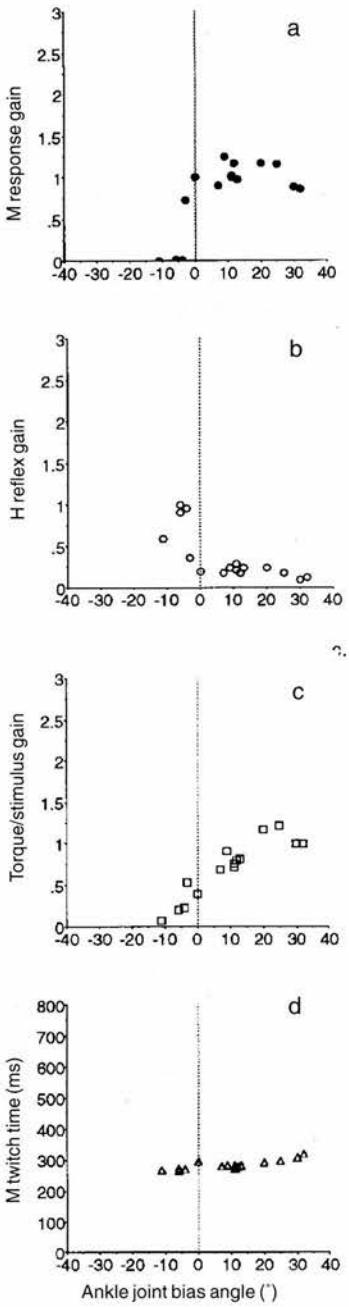

Figure 8: *Percutaneous axonal stimulation studies. M responses are compared with Hoffman reflex (H response) across joint range. (a) M response gain (% M amplitude/% stimulus amplitude) remains reasonably constant after dorsiflexion beyond neutral. (b) H reflex gain (% H amplitude/% stimulus amplitude) wanes after dorsiflexion beyond neutral. (c) % torque/% stimulus gain increases serially through almost the whole of dorsiflexion. (d) Twitch time (ms): this increases serially with increasing muscle stretch, irrespective of mode of muscular activation.*

ii) Torque gain

The reflex mechanical gain (normalised twitch torque/normalised tap torque) steadily rises, increasing threefold at +11° of dorsiflexion beyond neutral and then declining to original resting torques at +30° of dorsiflexion. It can be seen that the joint angle for maximum reflex mechanical gain differs from that of the maximum reflex EMG gain, the mechanical gain continuing to rise even though the EMG gain has started to wane. This indicates that there are mechanical factors which contribute to the reflex torque and which are capable of offsetting a drop in the number of reflexly recruited motor units (see electrical stimulation below).

iii) Twitch time

The twitch time remains constant at about 230 ms from resting plantar flexion to +10° of dorsiflexion beyond neutral and then steadily increases with further increments of dorsiflexion, reaching about 350 ms at +30°, corresponding to an absolute increase of 130 ms or 56% above that obtained in resting equinus. A muscle which has lengthened takes a little longer to contract, but the effect of muscle stretching prolongs relaxation especially.

iv) Twitch frequency

The inverse twitch time or predicted twitch frequency (not shown) declines from 4.25 Hz between −12° and +10° to 2.75 Hz with dorsiflexion to +30°. This is the same as saying that soleus muscle tetanisation would be more easily achieved in full dorsiflexion than at the resting angle.

Figure 7 shows the averaged EMG discharges at different joint angles in response to four percutaneous electrical stimulations of the soleus muscle via the posterior tibial nerve in the popliteal fossa (M response) compared with the electrically stimulated monosynaptic H responses and the accompanying muscle twitches (see Fig. 3a,b for explanation of M and H responses). At all joint angles the stimuli consisted of a fixed current of 56 mA with a pulse width of 100 μs. These electrically stimulated twitch data come from the same subject as in Figures 5 and 6 and were obtained at the same experimental session, so the conditions of electrical stimulation, including electrode placement, were similar to those for the tendon-tap reflex studies. Figure 8 illustrates these findings graphically. The soleus muscle is electrically stimulated at similar joint intervals to the tendon tap studies and the results can be summarised as follows.

i) The electrical stimulus artefact (Fig. 7) is present on the EMG trace as the first brief downward deflection. At −11° and −6° of plantar flexion, stimulation of the posterior tibial nerve in the popliteal fossa produces an H reflex EMG discharge some 30 ms later. From 0° onwards, the same electrical stimulus produces a large, direct EMG discharge (the M response), which arises a few milliseconds after the electrical stimulus (Figs. 7, 8a). This M response is seen as the largest deflection on the EMG traces from 0°, +9°, +13°, +20°, +25°, +30° of dorsiflexion; there is a small fall in amplitude of the M response between +10° and +30°. Some 30 ms after the M response is a small H reflex EMG discharge (Figs. 7, 8b) which is maximal at −5° of plantar flexion and diminishes serially with dorsiflexion beyond neutral. Although the muscle spindles are bypassed by the electrical stimulus, the diminishing H reflex indicates that reflex spinal alpha motor neuron recruitment is inhibited by muscle stretch.

ii) When the muscle is stimulated directly along the motor axon and independently of the spinal alpha motor neuron pool, the muscle twitch force (Fig. 8c) increases in strength to a maximum at +30°, indicating a mechanical advantage at near-maximum dorsiflexion for the soleus muscle. If the motor axons are stimulated directly, mechanical output of the soleus muscle increases with dorsiflexion through practically the whole joint range. This contrasts with the tap-induced reflex twitch force, which reaches a peak at +11° beyond neutral and declines thereafter.

iii) The muscle twitch time increases serially with each increment of dorsiflexion (Fig. 8d), apparently dependent on muscle length but independent of the mode of stimulation.

GROUP DATA

Figure 9 illustrates the effect of joint angle on the ankle jerk for nine healthy adults: nine right and eight left calves, each point representing the average of responses to four tendon taps. Figure 9a shows variation in normalised tendon tap torque across the joint range for the 17 adult soleus muscles. The group data confirm that the maximum reflex EMG gain occurs from −15° to −10° plantar flexion (Fig. 9b), in contrast to the reflex mechanical gain, which is maximal between 0° and +10° of dorsiflexion (Fig. 9c), indicating a net gain in reflex torque for any given tendon tap modulated by the joint angle. The angle for maximum reflex EMG gain differs from that of the resting joint angle, and the maximum reflex mechanical gain is obtained with a further dorsiflexion of 20° to 25°. Figure 9d shows the marked increase in the reflex twitch time for the group as a whole as the muscle is lengthened in increments from resting plantar flexion to maximum dorsiflexion, the regression equation indicating a mean of 265 ms at −25°, 267 ms at neutral, and 383 ms at +35° – a difference in twitch time of 118 ms across the joint range, or a 44.5% increase with dorsiflexion.

Figure 10 shows the reflex data for the soleus muscles of 22 children aged 3.9 to 13.6 years. The data represent data subsets from 41 soleus muscles, 22 right and 19 left respectively; one left limb was untested in a 3-year-old girl to shorten the test time, and in two cases no reflexes were obtainable on the left side after the right had been tested. These data from children are more scattered than those from adults. Nevertheless, the maximum reflex EMG gain is skewed to between −15° and −10° of plantar flexion (Fig. 10b) whereas the reflex torque gain is maximal between 0° and +10° curve (Fig. 10c). The reflex twitch time is likewise more scattered but increases with dorsiflexion, rising from a mean of 277 ms at −25° and 285 ms at neutral to 370 ms at +35° – a mean difference of 93 ms across the joint range, or a 33.5% increase. As Figure 4d illustrates, the slowest twitches occur in the youngest children, reaching the values for young adults by the age of 10 years, and apparently slowing again beyond the second decade. The reflex data for children and adults are similarly influenced by the joint angle.

These experiments indicate that short-term muscle stretch modulates reflex excitability, presumably by altering spinal motor neuron recruitment, demonstrating the effect of limb posture on the expression of the excitability of the nervous system.

Discussion

The effects of short-term passive changes in joint angle on stretch reflex output indicate the influence of peripheral factors

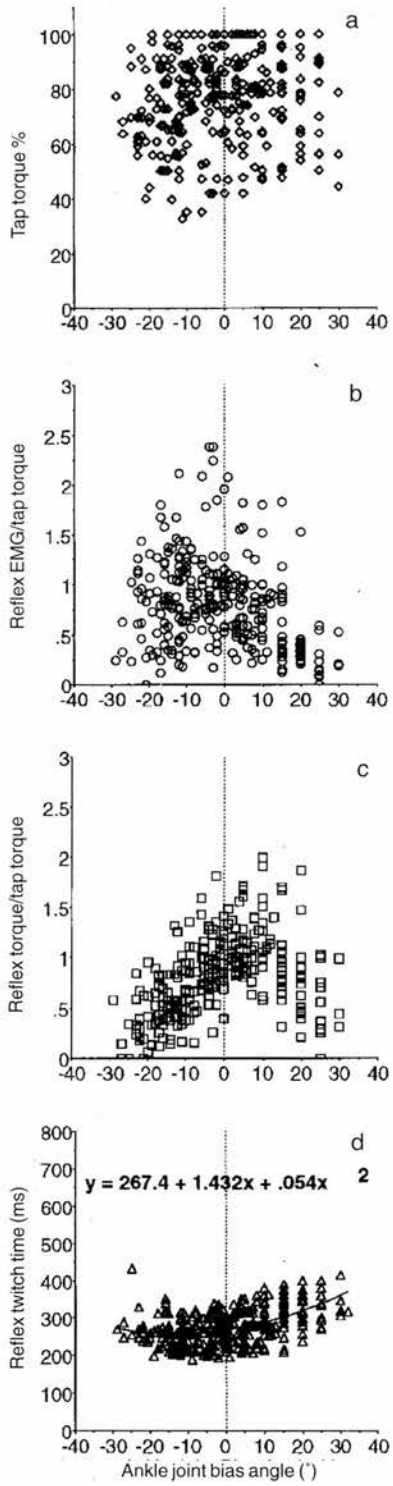

Figure 9: *Effect of joint angle on adult reflex excitability. (a) Normalised tendon tap stimulus (% tap torque) across joint range. (b) Reflex EMG gain (% reflex EMG/% tap torque) is maximal between −15° and −10° of plantarflexion. (c) Reflex torque gain (% reflex torque/% tap torque) is maximal between 0° and +10° of dorsiflexion. (d) Twitch time (ms) increases by 44.5% in maximal dorsiflexion. Nine healthy adults (6 males, 3 females): 9 right and 8 left legs. Each point represents an average of responses to four taps.*

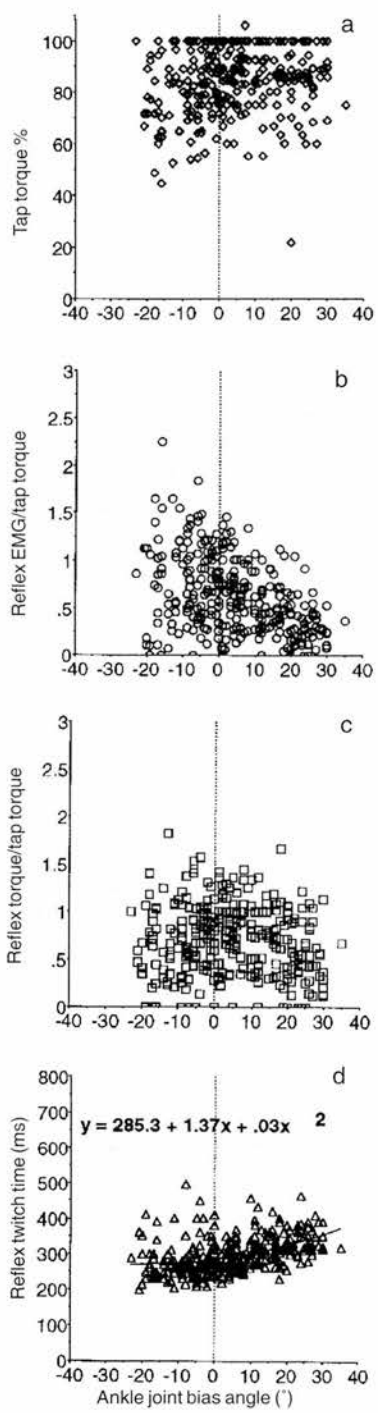

Figure 10: *Effect of joint angle on paediatric reflex excitability. (a) Normalised tendon tap stimulus (% tap torque). (b) Reflex EMG gain (% reflex EMG/% tap torque) is maximal in −10° to −5° plantarflexion. (c) Reflex torque gain (% reflex torque/% tap torque) is maximal between 0° and +10° of dorsiflexion. (d) Reflex twitch time (ms), increasing 35% with maximal dorsiflexion beyond neutral. Data are more scattered than for equivalent adult plots, possibly reflecting differences in muscle compliance with age (see Fig. 4b). Twenty-two children aged 3.9–13.6 years: 22 right and 19 left legs. Each point represents an average of responses to four taps.*

on this physiological phenomenon. This finding explains some of the physiological variability in eliciting and quantitating tendon jerks and velocity-dependent stretch reflexes within and between subjects as well as the large interobserver variations in the clinical setting. Much more interesting for motor physiology is the evidence for a *functional* neuromechanical joint range and of an optimal neuromechanical coupling in mild dorsiflexion beyond neutral, compared with an optimal neural angle of –15° to –10°.

While there is no doubt that a mechanical tap elicits a stretch reflex, the hammer blow varies from tap to tap, which is why the data from four consecutive taps were averaged: this practice was intended to average out the performance of the examiner as well as of the subject. If the change in tap-induced reflex EMG output had simply varied with the strength of the tendon taps, there would be no change in reflex EMG gain (ratio of the normalised reflex EMG to normalised tendon tap torque) over the joint range. This indicates a genuine gain in reflex motor unit output in mild plantar flexion beyond neutral, brought about by a change in muscle length which affects the recruitment of the spinal alpha motor neurons. In addition to modulating the tendon reflex, the joint angle has been shown to modulate the reflex EMG response to sinusoidal stretching in spastic quadriceps and hamstrings muscles in adults (Burke et al. 1971b) and children (Lin et al. 1994a).

It is clear from the present studies that the optimal joint angles for reflex EMG gain and reflex mechanical gain are out of phase, since the reflex mechanical gain achieves its maximum in mild dorsiflexion (optimal neuromechanical angle) beyond neutral, at an angle which is 20° to 25° more dorsiflexed than the angle of maximum reflex EMG gain (optimal neural angle). When the muscle was stimulated electrically via the posterior tibial nerve motor axons, the greatest motor torque arose close to the position of full muscle stretch, at +30° of dorsiflexion (a finding which agrees with the work of Sale et al. 1982), which we have called the optimal mechanical angle. What these data suggest is that the performance of the muscle cannot be assessed on the basis of the EMG output alone, since the mechanical output will vary according to the joint angle which influences that EMG output differently according to whether the muscle is directly or reflexly excited. This also means that direct axonal stimulation experiments to determine motor output provide only one picture of the way in which motor output varies with joint angle.

Accepting that the reflex alpha motor neuron output varies with the joint angle, is it the change in muscle length or in muscle tension which determines reflex excitability? Muscle tension is sensed by the tendon's Golgi organs, which are stimulated during active muscular contraction but are relatively quiescent in passive changes of muscle length (Houk and Henneman 1967). This means that the sudden, brief changes in tension produced by the tendon taps themselves are unlikely to be responsible for the changes in reflex excitability, nor are the passively sustained incremental changes in muscle tension produced by the bias torques, leaving either sensory afferents in the ankle joint capsule and soft tissues or, more likely, the muscle spindles themselves as the alternative candidates. In health, the spinal alpha motor neurons may be inhibited presynaptically by a number of influences, explored by Burke et al. (1971a) in a study in which the H reflex was inhibited by passive dorsiflexion. This inhibitory influence obtained when the Achilles tendon was stretched (altering its length) but not when it was squeezed (maintaining a constant length but stimulating sensory afferents). When the same investigators selectively blocked large Ia afferent fibres by inducing local ischaemia, the inhibitory effect of stretch persisted, indicating that type II (slow-conducting) small afferent fibres were responsible for the inhibition of stretch reflex excitability, such fibres forming the secondary spindle endings which are thought to monitor changes in muscle length (see also Burke and Lance 1973). The effects of calf ischaemia have been reported by Hultborn et al. (1996), who, unlike Burke and colleagues, found that 27 minutes of calf ischaemia abolished the inhibitory effects of previous dorsiflexion on the H reflex. However, Hultborn and colleagues were looking at the duration of the inhibitory influence of a previous, conditioning dorsiflexion.

The final reflex motor output is thus a summation of the effects of muscle length on neural output and on the output of the contractile elements intrinsic to muscles, and the effect of joint angle on the mechanical arrangements of the muscle–tendon–bony lever complex. This would explain why over the range of a few degrees in dorsiflexion, the reflex peak torque continues to rise despite the fact that the reflex EMG has started to wane: the mechanical advantage of stretch continues to produce a small increase in peak torque until, with further increments of dorsiflexion, the fall in the number of reflexly recruited alpha motor neurons is so great that it overrides any mechanical advantage: neuromechanical coupling becomes suboptimal. The optimal angle for neuromechanical output is neither the resting angle nor the extreme of dorsiflexion, but midway between the two. Sale et al. (1982) discuss the fact that the calcaneal lever arm (defined as the shortest distance between the line of pull of the Achilles tendon at its insertion and the talotibial joint) diminishes by two-thirds in full dorsiflexion from a maximum in plantar flexion. If Fp is the intramuscular force in full plantar flexion and Lp the length of the lever arm, the torque T generated will be $T=Fp \times Lp$. From this the relative theoretical intramuscular force (Fd) necessary to produce an equivalent torque in full dorsiflexion can be calculated. Assuming the dorsiflexed lever arm to be $Ld=^2/_3Lp$, $T=Fd \times ^2/_3 . Lp=Fp \times Lp$, giving $Fd=^3/_2Fp$. This means that for any arbitrary soleus torque in full plantar flexion to be matched by an equal torque in the dorsiflexed (stretched) soleus, the dorsiflexed muscle must actually be capable of generating 1.5 times the plantar flexed intramuscular force.

Therefore, the mechanical leverage helps to offset a reduction in intramuscular force generation of the plantar flexed soleus.

Current texts on basic muscle function (see Moseley 1992, p 61; Rab 1993, p 109–10) continue to stress that the muscle achieves its maximum force at its *resting length*, this being based on the 'Blix curve', which in turn is derived from in vitro experiments in which denervated muscles are held between two fixed points in a dish. This Blix model departs from the data presented here and from those of Marsh et al. (1981) and Sale et al. (1982), in which the torque generation by the soleus muscle at resting plantar flexion is negligible, and resting plantar flexion corresponds to the physiological resting length of the muscle–tendon complex. The significance of optimal neuromechanical coupling for normal physiology can be illustrated by a few examples.

1) In dynamic gait, during the single support phase (stance), the calf muscles act eccentrically to decelerate the tibia as it

rotates forwards at the ankle between the first and third rocker phases. Sutherland and colleagues (1980) demonstrated that during the single support phase, 'ankle dorsiflexion was first resisted, then arrested and reversed by plantar flexor muscles acting *eccentrically*' and this eccentric activity occurs between –5° and +10° of the joint range about the neutral angle – precisely the joint interval across which neuromechanical coupling was maximal in our study. As stated above, numerous studies of H reflex modulation during gait have now confirmed the functional importance of this joint range. A practical example of a disturbance of this relationship is the deleterious effects of heel-cord overlengthening. Under these circumstances the new 'optimal' neuromechanical angle occurs at a more dorsiflexed position, so that the soleus can no longer eccentrically decelerate the shank as it rotates forwards at the ankle, all of which produces the well-known crouch gait.

2) For innervated muscle, control of the force output is proportional to the number (and size) of the motor units participating in active muscle contraction (the Henneman size principle; see Henneman et al. 1974). We have previously shown that the most forceful, rapid, and convenient voluntary alternating movements at the ankle occur close to the neutral joint angle and appear to involve the largest and fastest motor units (Lin et al. 1996a, Lin 1997), indicating that certain postures favour the optimal execution of voluntary motor tasks, the amplitude of any alternating movements varying inversely with the square of the frequency of the motion. Accordingly, large joint excursions can only be performed slowly and the fastest movements have small amplitudes, oscillating close to the optimal neuromechanical angle. It has been hypothesised that children may learn to use favourable postures to improve their dexterity in the performance of rapid alternating movements (Lin 1997). As indicated in this study, reflex motor output in resting plantar flexion is weak and brief, whereas in extreme dorsiflexion it is weak and up to 45% slower than that of the resting angle. Although maximum voluntary contraction appears to override the joint angle modulation of motor neuron output, as in running (Capaday and Stein 1987), few activities are regularly performed in conditions of maximum voluntary motor drive, and the effect of the joint angle at lower levels of motor activity may be of functional significance in producing economical motor strategies. It is likely that the spinal alpha motor neuron pool undergoes modulation during physiological movements, being relatively refractory to recruitment at the extremes of the joint range, except at the expense of greater descending motor drive.

3) Involuntary alternating movements such as ankle clonus are likely to be influenced by the joint angle, the extremes of the joint angle being refractory to clonus because of low neuromechanical output, the strongest beats occurring just beyond neutral, becoming slower and weaker with progressive dorsiflexion as the reflex EMG and reflex torque dwindle and the muscular twitches get slower. The clonic phenomena are likely to change after peripheral interventions such as tendon lengthening or plaster immobilisation at an increased muscle length, which would shift the excitability curve to the right as well as have an effect on the contractile properties of the muscle, i.e. alter fibre-type composition. These predictions have beeen explored elsewhere (J-P Lin, JK Brown, and EG Walsh, in preparation).

These findings are of relevance to current orthopaedic practice, sports medicine, rehabilitation, and associated physical therapies, in focusing attention on how this reflex modulation might influence motor function in states of health, injury, and disease, and how interventions may produce a new motor picture which can be partly predicted from this model for joint angle.

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